Fibro-Osseous Lesions of Cranio-Facial Complex

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Abstract

Fibro-osseous lesions (FOLs) of the craniofacial complex are represented by a variety of disease processes that are characterized by pathologic ossifications and calcifications in association with a hypercellular fibroblastic marrow element and share microscopic features. The definitive diagnosis can rarely be rendered on the basis of histopathologic features alone; rather, procurement of a final diagnosis is usually dependent upon assessment of microscopic, clinical and imaging features together. Fibrous dysplasia, ossifying fibroma, and osteoma are three distinct entities that lie along a continuum from the least to the most bony content. They have similar appearance and makeup; however, their clinical implication vary further complicate matters, a number of other disease processes demonstrate clinical, radiographic, and microscopic features that bear resemblance to those encountered in recognized fibro-osseous conditions. The objective of this article is to review the most current clinicopathologic, radiographic, and molecular studies of FOLs to aid the surgical pathologist in the recognition and diagnosis of this diverse group of maxillofacial lesions.

Keywords: Fibro-Osseous Lesions; Cranio-Facial Fibrous Dysplasia; Ossifying Fibroma; Osteoma

Introduction

Fibro-osseous lesions (FOLs) of the craniofacial complex are represented by a variety of disease processes that are characterized by pathologic ossifications and calcifications in association with a hypercellular fibroblastic marrow element and share microscopic features [1]. Whereas some are diagnosable histologically, most require a combined assessment of clinical, microscopic and radiologic features. In turn, some fibro-osseous lesions (FOL) of the craniofacial complex are unique to that location whereas others are encountered in bones from other regions [2]. Reactive, neoplastic, developmental and dysplastic pathologic processes are subsumed under the rubric of FOL and treatment varies from disease to disease.

FOLs can involve paranasal sinuses, skull base, maxillofacial region. FOL involving skull base and paranasal sinuses are uncommon. Fibrous dysplasia, ossifying fibroma, and osteoma are three distinct entities that lie along a continuum from the least to the most bony content. They have similar appearance and makeup; however, their clinical implication vary [3].

Whereas some investigators include giant cell lesions of bone with FOL, lesions of this nature will not be included in this study with the exception of the trabecular variant of ossifying fibroma which is essentially a FOL yet may contain foci of multinucleated giant cells [4].

Fibrous dysplasia is a benign dysplastic process of altered osteogenesis that may occur within a single bone (monostotic) or multiple bones (polyostotic) [5]. When polyostotic fibro-osseous lesions typical for fibrous dysplasia are associated with other anomalies and endocrinopathy, this variant form constitutes the McCune-Albright syndrome (MAS).

Monostotic fibrous dysplasia of the craniofacial complex is often confused with other FOL, typically ossifying fibroma and diffuse sclerotic osteomyelitis of the mandible, diseases that manifest unique clinicoradiologic features. Monostotic fibrous dysplasia occurs in the jaw, frontal, ethmoidal, temporal and calvarial bones [5]. In the less prevalent polyostotic fibrous dysplasia (PFD) and McCune-Albright syndrome (MAS), the craniofacial region is involved in 90% of the cases and the anterior cranial base is involved in over 95% of cases (Figure 1) [6].

![Figure 1: Extensive fibrous dysplasia involvement of the cranial base in a patient with MAS [6].](image)

Depending on the type and location of FD, the signs and symptoms vary and include facial deformity and asymmetry, vision changes, hearing impairment, nasal congestion and/or obstruction, pain, paraesthesia, and malocclusion. Many patients are asymptomatic and the diagnosis is made when a family member, friend or health care provider who has not seen the patient for a period of time notices asymmetry, or there is an incidental abnormality noted on dental or panoramic x-rays or on a head and neck computed tomogram (CT).

When rapid enlargement occurs, adjacent vital structures, such as the optic nerve, globe and auditory canal/structures and nasal airway may be invaded or compressed, resulting in functional deficits. For these reasons, some authors have advocated aggressive surgical resection to avoid potential blindness or hearing loss [7]. Rapid enlargement of FD in the nasal bones, maxilla or mandibular symphysis may result in airway obstruction by obliteration of the nasal cavity or by posterior displacement of the tongue. However, it has recently been demonstrated that such aggressive behaviour with rapid expansion is the exception and that a conservative expectant approach is more prudent [8].

A thorough history and physical examination are necessary to determine the extent of disease and to determine whether the FD is isolated or one of multiple lesions associated with PFD or MAS.

Documentation of the onset and types of symptoms, presence of functional impairments and duration are imperative. Inquiries should include onset of menarche in females (to rule-out precocious puberty), other endocrine abnormalities or pathologies (such as hyperthyroidism, pituitary abnormalities, and renal phosphate wasting), growth abnormalities (review of growth charts), and history of fractures (to rule-out the presence of other FD lesions in the extremities) as well as the presence of skin lesions (café-au-lait lesions). These questions are particularly critical in young patients where underlying endocrine abnormalities may not have been detected and aggressive management is warranted. If there are any positive responses to the above inquiries, a referral to an endocrinologist is strongly recom
mended to rule out PFD or MAS. A skeletal survey or bone scan may be indicated if there is a suspicion of PFD or MAS, particularly in a patient that is not skeletally mature. Additional FD lesions beyond the craniofacial region require further evaluation by an orthopaedic surgeon [6].

Asymmetry and swelling are the most common complaints when FD is found in the bones of the facial skeleton. Secondary deformities due to slow growing FD include vertical dystopia (difference in the vertical position of the eyes), proptosis, frontal bossing, facial and jaw asymmetries or canting (angular deviation from a vertical or horizontal plane). The degree of facial deformity varies, but those with MAS are the most severely affected, particularly when associated with untreated or inadequately treated growth hormone excess [9].

Annual evaluations may be adequate. The patient’s concerns and symptoms, clinical assessment including sensory nerve testing in the region of involvement, photographs, and facial CT should be obtained at each visit. An annual CT may be necessary for the first 2 years; however, the interval may be lengthened based on the clinical findings. Surgical contouring by a maxillofacial or craniofacial surgeon is indicated if the patient is bothered by facial disfigurement [10].

In patients with non-aggressive but active FD, it is ideal to wait until the lesion becomes quiescent and the patient has reached skeletal maturity before performing an operation. However, in cases where the patient’s psychosocial development may be impaired due to the facial deformity, surgical contouring and/or resection may be warranted. The patient and family must be aware of the potential for regrowth if the lesion cannot be resected completely, which is often the case. In cases of PFD or MAS where the disease is extensive, the lesions are often not respectable. Repeat surgical contouring and extensive debulking may be necessary to achieve acceptable facial proportions [11].

In the future, improvement in CT imaging and software will allow for accurate surgical simulation and intraoperative navigational tools may guide the surgeon throughout the contouring. Advanced CT software is useful for superimposition of pre- and post-operative images. These can then be compared to follow-up CT scans to determine stability of the result or the presence of regrowth. Despite these new imaging technologies, there is no therapy or technology that can predict and/or prevent regrowth.

Malignant transformation of FD has been reported in less than 1% of cases of FD [12]. Typically, the malignancy is a sarcomatous lesion, most often osteosarcoma but fibrosarcoma, chondrosarcoma, and malignant fibrohistiocytoma have also been reported [13]. The diagnosis may be difficult, particularly in cases of low-grade osteosarcoma [14]. The treatment is based on the management of the malignancy and resection with adequate margins is necessary.

The most common radiographic characteristic of craniofacial FD is a “ground-glass” appearance with a thin cortex and without distinct borders [15]. In an ongoing study at National Institute of Health (NIH), [16] it was demonstrated that the typical characteristics of FD on CT and the natural radiographic progression may vary from a “ground-glass” or homogenous appearance to a mixed radio-dense/radio-lucent lesion as the patient ages in pre-pubertal patients with PFD or MAS, the lesions most often appear as homogenous, radio-dense lesions on CT. As these patients enter the second decade of life, the FD lesions progress to a mixed appearance, which stabilizes in adulthood but does not resume a homogenous appearance. While the change to a mixed radiographic appearance alone does not require further biopsy or investigation, we recommend careful monitoring and intermittent craniofacial CT during the pubertal phase of the young patient. This period of change in CT appearance coincides with case reports of increased activity of the FD lesions either through rapid growth, worsening facial asymmetry, malignant transformation, or association with other pathologic, radiolucent lesions such as aneurysmal bone cyst (ABC) and accelerated expansion [17] (Figure 2) [6].

Figure 2: Variations in CT appearance of fibrous dysplasia based on age. A) FD in the young patient most often appears as homogenous, radiodense lesions often described as having a ground glass appearance on CT. B) As these patients enter adolescence, the FD lesions progress to a mixed appearance which stabilizes in adulthood (C) but does not necessarily resume a homogenous appearance [6].

The expansion of the external surface of the affected bone assumes a more grotesque, but still recognizable shape whereas the internal surfaces expand into orbital, nasal and sinus cavities, fissures, fossae and neural and vascular canals. The lesion, if large often nearly completely obliterates the maxillary sinus (Figure 3) [18]. The above pattern is altered if the FD undergoes cystic degeneration with formation of a large aneurysmal bone cyst. Then the affected part of the lesion may lose its anatomical shape and becomes spherical (Ferretti and co-authors’ report) [19].

Figure 3: A bone window of a coronal CT section of FD affecting the maxilla. It obliterates almost completely the antral cavity. Although the lesion displays a generally homogenous ground glass appearance, there are areas that display both cotton wool and peau d’orange features. It extends into the hard palate, the alveolar process to envelop the molars’ roots and the zygomatic arch (it has also secondarily envelopes the arch’s medial surface). It displaces the inferior lateral wall of the nasal cavity medially. It also displaces the buccal (lateral) aspect of the maxilla laterally [19].

A bone biopsy, by the appropriate surgical specialist, should be obtained to confirm the diagnosis of FD, if the site is amenable to biopsy. Unfortunately, the histology does not predict the biological behaviour of these lesions [7]. Biopsy of FD does not specifically induce growth of the lesion. However, FD lesions may be quite vascular and bleeding can be brisk. The surgeon should be prepared to deal with this. If the lesion is quiescent or asymptomatic, and/or in the cranial base, a biopsy may not be possible or necessary. History, clinical examination and the classic radiographic presentation are often adequate to establish the diagnosis of FD.

Fibrous dysplasia can affect sinuses, teeth, orbit/sphenoid bone, auditory canal/temporal bone. The sinuses may be affected by FD, with the most frequent site being the sphenoid sinus, followed by the ethmoid and maxillary sinuses [20] (Figure 4) [6].

The entire sinus can be completely obliterated by FD, yet surprisingly the incidence of sinusitis is not greater than the general population in these patients. This may be explained by the loss of air space and Schneiderian membrane in an obliterated sinus and the elimination of a source of infection. Patients typically complain of nasal congestion (> 34% of those with symptoms and sinus involvement), headaches or facial pain, recurrent sinusitis, and hyposmia. This appears to be associated with FD in the inferior turbinate and the subsequent hypertrophy.

Teeth the most common anomalies included: tooth rotation, oligodontia (congenital condition in which some of the teeth are missing), displacement, enamel hypoplasia, enamel hypo mineralization, taurodontism (is a condition found in the molar teeth of humans whereby the body of the tooth and pulp chamber is enlarged vertically at the expense of the roots. As a result, the floor of the pulp and the furcation (bone loss) of the tooth is moved apically down the root), retained deciduous teeth, and attrition. (is the loss of teeth structure by mechanical forces from opposing teeth. Attrition initially affects the enamel and, if unchecked, may proceed to the underlying dentin. Once past the enamel, attrition quickly destroys the softer dentin. Erosion is a very important contributing factor to the loss of tooth substance by attrition) [6].
Common findings associated with PFD around the eye include proptosis, dystopia, and hypertelorism due to the involvement of the frontal, sphenoid, and ethmoid regions. Less common findings include: optic neuropathy, strabismus, lid closure problems, nasolacrimal duct obstruction and tearing, trigeminal neuralgia and muscle palsy with skull base involvement [21].

The temporal bone is frequently involved (> 70%) in patients with craniofacial PFD or MAS [22], while temporal bone involvement is uncommon in monostotic disease [23]. In a recent analysis by DeKlotz., et al. [22] despite the high incidence of disease of the temporal bone in PFD, nearly 85% of patients had normal or near-normal hearing; 10% had conductive hearing loss due to PFD, approximately 4% had sensorineural or mixed hearing loss (both conductive and sensorineural), and the remainder had hearing loss due to other, non-PFD related causes [19]. In most cases, the degree of hearing loss was mild (77%) and did not correlate to the amount of disease involvement of the temporal bone. The common causes of hearing loss appeared to be narrowing of the external auditory canal due to the surrounding FD (Figure 5) [6] and fixation of the ossicles within the epitympanum from adjacent involved bone (Figure 6) [6].

**Figure 5:** Narrowing of the external auditory canal due to fibrous dysplasia (FD). A) A CT image of an axial slice through the temporal bone shows a narrowed external auditory canal (arrow) B) Narrowing of the canal is shown and can be compared to a normal canal in (C) [6].
Fibrous dysplasia of the cranio-facial complex may also be associated with soft tissue myxomas, the Mazabraud syndrome is a rare syndrome comprising of:

- fibrous dysplasia: usually polyostotic
- multiple soft tissue (intramuscular) myxomas: typically, in large muscle groups it is most frequently seen in women (~70%) and usually present in middle age (mean age 46; range 17 - 82) There is increased risk of osseous malignant transformation [24].

**Ossifying Fibromas**

These are neoplasms in the true sense, exhibiting progressive proliferative capabilities with bony expansion and, importantly, well defined margins radiologically. Subsumed under this diagnostic category are ossifying/cementifying fibroma, not otherwise specified (NOS), implying that the clinicopathologic features do not conform to the other types of ossifying fibromas; these specific subtypes include psammomatoid variant of ossifying fibroma, trabecular variant of ossifying fibroma and gigantiform cementoma, the latter of which may show an autosomal dominant genetic or “familial” underpinning.

Most ossifying fibromas are single focal lesions; however, gigantiform cementoma is typically multifocal and may occur in all four jaw quadrants in a single patient. There are also reports of lesion multiplicity in the other forms of ossifying fibroma yet such occurrences are quite rare.

The most common form of ossifying fibroma occurs in the maxilla and mandible and represents a neoplastic process that is typically painless, presenting with expansion of the buccal and lingual cortices and in larger lesions expand the inferior aspect of the mandible [25]. Teeth in the vicinity of these lesions are often displaced superiorly (Mandibular lesions) or inferiorly (Maxillary lesions) and the latter typically expand into the maxillary antrum. Radiographically, early stage lesions in their formative phase, are typically radiolucent since the osseous element is non-calcified osteoid. Over time, the tumors become progressively radiopaque as more matrix calcifies (Figure 7) [4].
Figure 7: Ossifying fibroma NOS. Radiographically, the lesions are and radiolucent with internal floccular to dense opacities [4].

Juvenile Ossifying Fibroma the term juvenile ossifying fibroma (JOF) also known as Juvenile Active Ossifying Fibroma [26] and juvenile aggressive ossifying fibromas is used in the literature to describe two distinct clinicopathologic entities; (1) Trabecular juvenile ossifying fibroma (TrJOF) and (2) Psammomatoid juvenile ossifying fibroma (PsJOF) [27].

Trabecular Juvenile Ossifying Fibroma TrJOF also known as trabecular desmo-osteoblastoma is defined as a lesion affecting the jaws of children composed of a cell rich fibrous stroma containing bundles of cellular osteoid and bone trabeculae without osteoblastic rimming, and aggregates of giant cells [28].

The maxilla and the mandible are the dominant sites of incidence. Occurrence in the maxilla is slightly more frequent than in the mandible. Origin in extra gnathic locations is extremely rare. Clinically, TrJOF is often characterized by a progressive and sometimes rapid expansion of the affected area; pain is a rare symptom [29].

Radiographically, the tumor is expansive and may be fairly well demarcated, with cortical thinning and perforation. Depending on the amount of calcified tissue produced, the lesion will show varying degrees of radiolucency or radiodensity. Ground-glass as well as a multilocular honeycomb appearance has been described (Figure 8) [4].

Unlike TrJOF, psammomatoid juvenile ossifying fibroma (PsJOF) is a lesion that affects predominantly the extragnathic craniofacial bones, particularly centered on the periorbital, frontal, and ethmoid bones [26]. PsJOF was initially described by Margo, in 1985 [30] described PsJOF as a distinctive solitary fibro-osseous lesion of young individuals that affects the orbit and shows distinguishing histologic features.

PsJOF was also reported under the designation juvenile active ossifying fibroma by Johnson, et al. [31] and juvenile ossifying fibroma with psammoma like ossicles [28] and psammous desmo-osteoblastoma by Makek [26]. PsJOF is not classified under osseous tumors of the jaws in the Armed Forces Institute of Pathology (AFIP) Atlas of Tumor Pathology [32] nor was it considered a gnathic tumor in the WHO histologic classification of odontogenic tumors [33].

PsJOF is clinically manifested as bone expansion that may involve the orbital or the nasal bones and sinuses. Orbital extension of sino-nasal tumors may result in proptosis, and visual complaints including blindness, nasal obstruction, ptosis, papilledema, and disturbances in ocular mobility.

Radiographic examination shows a round, well-defined, sometimes corticated osteolytic lesion with a cystic appearance [34]. Sclerotic changes are evident in the lesion which may show a ground-glass appearance. In computed tomographic scans set on bone window, the lesions appear less dense than normal bone (Figure 9) [4]. The lesions may range in size from 2 to 8 cm in diameter. PSOF may appear multiloculated on CT scans. Areas of low CT density may be noted due to cystic changes. It is stated that in the facial skeleton a well-circumscribed expansive mass with a thick wall of bone density on CT scan and enhancement of this area on post contrast MR image is strongly suggestive of psammomatoid juvenile ossifying fibroma.
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Figure 9: Aggressive Ossifying Fibroma, Psammomatoid Type. Paraorbital well demarcated expansile radiopacity [4].

Osteoma

An osteoma is a benign osteogenic tumor characterized by compact or cancellous bone proliferation. It may be classified as peripheral, central, or extra skeletal. A peripheral osteoma arises from the periosteum, a central osteoma from the endosteum, and an extra skeletal osteoma in the soft tissue [35]. The pathogenesis of osteomas is not completely known. They are referred to developmental anomalies, true neoplasms, or reactive lesions triggered by trauma, muscle traction, or infection [36].

Osteomas are found mainly in the craniofacial bones. A peripheral osteoma (PO) occurs most frequently in the paranasal sinuses. Other locations include the orbital wall, temporal bone, pterygoid processes, and external ear canal [37]. The most frequent sites affected in the mandible are the posterior body, followed by the condyle, angle, ascending ramus, coronoid process, anterior body, and sigmoid notch [38]. It has been reported that osteomas can occur at any age and that males and females are equally affected [39].

Peripheral osteomas are slow-growing lesions and, clinically, they usually remain asymptomatic. However, when they reach a large size, they can produce swelling and asymmetry.

The frontal-ethmoidal sinus is the most frequent site in the paranasal sinuses and even so majority of these are micro lesions averaging about 5 mm in size and hence are usually asymptomatic [40]. Larger lesions, those up to 30 mm in diameter, are still more infrequent, but are more symptomatic by causing varying nasal/paranasal sinus inflammatory/infective and obstructive symptoms, and occasional intracranial/orbital complications [41].

Giant fronto-ethmoidal osteomas, lesions larger than 60 mm, are very rare indeed; [42] they usually have wide based attachments to craniofacial bones, and extensive involvement of the anterior skull base and the orbital-nasal complex. Surgical excision of such lesions is therefore more engaging, usually involves extensive craniofacial soft tissue and bony dissection and post lesionectomy reconstruction of the surgical dissection field with modern-day advanced microsurgical tools and techniques [43].

 Patients with osteomas should be evaluated for Gardner’s syndrome (GS). This syndrome is an autosomal dominant disease characterized by gastrointestinal polyps, multiple osteomas, skin and soft tissue tumors, and multiple impacted or supernumerary teeth. Intestinal polyps are predominantly adenomas and may progress to malignancy in almost 100% of patients [45]. Because the osteomas may be seen in the earlier stage of GS, the dentists may play an important role in the diagnosis of colonic polyposis.

The exact etiology and pathogenesis of peripheral osteoma is unknown. Neoplastic and reactive causes have been suggested as possible etiologic factors. Kaplan, et al [39] and Woldenberg, et al [46] suggested that some peripheral osteomas may be reactive rather than neoplasms, probably associated with trauma. Also, some authors have reported that as many of the peripheral osteomas are located on the lower border of the mandible, it is possible that muscle traction plays a role in the development of peripheral osteomas [46].

Clinically, peripheral osteoma appears as a unilateral and well-circumscribed mass ranging from 10 to 40 mm in diameter [47]. Lesions are usually asymptomatic and can be discovered in routine clinical and radiographic examination. Sometimes, depending on the location and size of the lesion, it may cause swelling, facial asymmetry, and functional impairment [48]. The swelling is usually painless.

The CT is the best imaging modality for determining the location and real extension of the lesion [46]. Peripheral osteomas, in most cases, are easy to recognize because of their classic radiographic findings. On radiological imaging, a peripheral osteoma of the mandible is a classically well-circumscribed, round or oval, mushroom-like radiopaque mass with distinct borders [49]. The lesion may be sessile and attached to the cortical plates with a broad base. If a peripheral osteoma is pedunculated, a narrow contact area can be seen between the lesion and the compact bone.

Differential diagnosis: peripheral osteoma should be differentiated from several pathologic entities, such as exostoses, osteoblastoma, and osteoid osteoma, late-stage central ossifying fibroma, or complex odontoma. Exostoses are bony excrescences that usually stop growing after puberty, differentiating them from osteomas [50]. The borders of central ossifying fibromas are well-defined, and a thin, radiolucent line may separate it from the surrounding bone. A sclerotic border may be present in the bone next to the lesion [51].

Osteoblastomas and osteoid osteomas are more frequently painful and grow more rapidly than peripheral osteomas [37]. A complex odontoma presents as a well-defined radiopacity situated in bone, but with a density that is greater than bone and equal to or greater than that of a tooth. It is also surrounded by a narrow radiolucent rim [51].

**Figure 11:** Panoramic radiograph showing a solitary, round, well-defined radio-opaque mass without a radiolucent rim on the left side of the body of the mandible. The lesion extended distally of the second premolar till the mesial aspect of the second molar distal root [52].

**Figure 12:** Coronal Computed tomography (CT) of the same case above showing a large, well-circumscribed, pedunculated mass attached to the buccal surface of the left mandibular body. Three-dimensional reconstruction image showing localization and extension of the lesion [52].
Fibro-Osseous Lesions of Cranio-Facial Complex

**Aim of the Work**

The study aims to assess the role of multidetector CT in assessment of fibro-osseous lesions of the craniofacial complex.

**Materials and Methods**

This study includes 25 patients. Their age ranges from 15 - 64 years old with a mean age 40 +/- 35 years. All the studied individuals were chosen selectively regarding complain (those with known fibrous dysplasia, or facial disfigurement or facial swelling) regardless of age and gender. Cases were referred to radiology department of Alexandria Main University Hospital, in the period between October 2012 till May 2013.

All the studied patients were subjected to the following:

- The medical ethics were considered: the patients should be aware of examination, patient approval was obtained, the economic status of the patient was considered and the patient got benefit from examination.
- Informed consent was written from all subjects and in cases of children their parents gave the consent to the investigation in written form
- Full history taking.
- Full clinical examination especially to the head and neck.
- Plain x-ray
- MDCT.
- 3D reconstructive images.

The present series studied 25 patients. The examinations were carried out in the following sequence:

Patients were instructed to minimize head movement during examination as much as possible in order to minimize motion artifacts.

Instructions were also given to remove any prohibited materials as hairpins. All examinations were performed on a multislice CT (MSCT) scanner (emotions 6, Siemens, Germany) six row unit or General Electric Light Speed CT scanner (emotions 4, USA) four row unit.

The patient was positioned supine on the table, axial scans were obtained through the entire skull from frontal sinus upward to lower border of mandible downward considered as landmarks. Axial volumetric acquisition was taken and completed with reformations of coronal and sagittal images. Axial sections were made parallel to the infraorbital-meatal line from upper border of frontal sinus down to the end of the mandible. MDCT technique generally consists of thin-collimation (0.6 mm) scanning performed with a 0.3 mm overlap, an advanced table 1.3 mm/rotation, a voltage of 120 KV with an intensity of 200 mA.

Axial images were reconstructed at 0.6 mm thickness and with a reconstruction increment of 0.3 mm resulting in more than 200 2D images to reduces stair-step artifacts using a high resolution bone algorithm. Matrix size was 512 x 512 in General Electric Light Speed (emotions 4, USA). Coronal sections were reconstructed as nearly perpendicular to the axial plane as possible. The images were displayed on a head field of view, window width of 350 HU for soft-tissue structures and window center of 700 HU and window width of 3500 HU for bone structures.

All cases were examined in all the three orthogonal views namely dead axial, dead sagittal and coronal view perpendicular to the hard palate and scanned for any variations in their normal anatomy. Any suspicious variation identified on a particular plane is pinpointed with the crosshair option (found in most imaging software) in other planes for proper assessment. This was particularly handy feature in cases when the identification of a certain variation was inconclusive in one certain plane. Being able to examine the same exact location in the other plane gives a broader understanding and imagination of the three-dimensional orientation of that particular position.

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The MDCT images were reconstructed using filtered reconstruction algorithms. The thin slices were sent to the workstations General Electric Light Speed advantage 4.2, after reconstruction of the raw data using sharp bony (B70) and smooth soft tissue reconstruction algorithms (B30), where they were available to view in axial, sagittal and coronal planes.

The images were used in 3D reconstruction volume rendering with Image J software (syngo) where it was taken in multiple angles with bone and soft tissue windows for further clarification and for planning for surgical interference.

Results

The present work included 25 patients with fibro-osseous lesions of the craniofacial complex. They were 10 males and 15 females. Their ages ranged between 15 and 60 years with a mean age of 40 +/- 35 years as shown in Table 1.

<table>
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<th>Males (%)</th>
<th>Females (%)</th>
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*Table 1: Distribution of patients according to age and sex.*

All patients considered the examination acceptable, and no discomfort or complications were found by caretakers. In the present study, the cranio-facial fibrous dysplasia represents almost half of the presented cases (48%) followed by osteomas then ossifying fibroma as shown in Table 2.

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<th>Final diagnosis</th>
<th>Number of patients</th>
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<td>48</td>
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<tr>
<td>Osteoma</td>
<td>9</td>
<td>36</td>
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<tr>
<td>Ossifying fibroma</td>
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<td>Total</td>
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*Table 2: Distribution of all patients according to final diagnosis.*

Fibrous Dysplasia

Regarding the sex distribution, two thirds of patients (8 patients out of 12) with fibrous dysplasia was diagnosed more frequently among females (66.6%) in the present study and half of the patients (6 patients out of 12) with fibrous dysplasia were in their 5th decade as shown in Table 3.

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<tr>
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Table 3: Distribution of the patients with fibrous dysplasia (n = 12) according to the age and sex.

Regarding complaint in the present study, twelve patients were diagnosed to have cranio-facial fibrous dysplasia. Two third of patients (8 patients out of 12) were presented with headache (66.6%) while only two patients were presented with facial deformity (16.6%) and 4 patients out of 12 were presented by facial swelling (33.3%) as shown in Table 4.

<table>
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<th>Complaint</th>
<th>Number of patients</th>
<th>% out of patients with fibrous dysplasia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>8</td>
<td>66.6</td>
</tr>
<tr>
<td>Swelling</td>
<td>4</td>
<td>33.3</td>
</tr>
<tr>
<td>Facial deformity</td>
<td>2</td>
<td>16.6</td>
</tr>
</tbody>
</table>

Table 4: Distribution of the patients with fibrous dysplasia (n = 12 patients) according to complaint.

Regarding the site of implication of the cranio-facial fibrous dysplasia, the maxillary bone and sinuses mainly ethmoid, sphenoid, and maxillary are most frequently affected (4 patients out of 12) (33.3%) each, followed by equal distribution for the mandibula, temporal bones and orbit (2 patients out of 12) (16.6%) each. The skull base, zygoma and occipital condyle are less frequently affected (only one patient each) (8.3%) each in the present study as shown in Table 5.

<table>
<thead>
<tr>
<th>Region</th>
<th>Number of patients</th>
<th>% out of patients with fibrous dysplasia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maxilla</td>
<td>4</td>
<td>33.33</td>
</tr>
<tr>
<td>Mandible</td>
<td>2</td>
<td>16.66</td>
</tr>
<tr>
<td>Sinuses</td>
<td>4</td>
<td>33.33</td>
</tr>
<tr>
<td>Skull base</td>
<td>1</td>
<td>8.33</td>
</tr>
<tr>
<td>Temporal bone</td>
<td>2</td>
<td>16.66</td>
</tr>
<tr>
<td>Occipital condyle</td>
<td>1</td>
<td>8.33</td>
</tr>
<tr>
<td>Zygoma</td>
<td>1</td>
<td>8.33</td>
</tr>
<tr>
<td>orbit</td>
<td>2</td>
<td>16.66</td>
</tr>
</tbody>
</table>

Table 5: Distribution of the patients with fibrous dysplasia according to the site of implication.

Regarding the involvement of bones, the number of patients were equal for both single bone involvement (monostatic 50%) and multiple bone involvement (polyostotic 50%) as shown in Table 6.

<table>
<thead>
<tr>
<th>Feature</th>
<th>Number of patients</th>
<th>% out of patients with fibrous dysplasia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polystotic</td>
<td>6</td>
<td>50</td>
</tr>
<tr>
<td>Monostotic</td>
<td>6</td>
<td>50</td>
</tr>
</tbody>
</table>

*Table 6: Distribution of patients (n = 12 patients) according to bones involved.*

None of the patients with polyostotic fibrous dysplasia showed features of McCune-Albright syndrome. According to the laterality of the lesion, the fibrous dysplasia was unilateral and specifically affecting the right side in most of the cases (11 patients out of 12) as shown in Table 7.

<table>
<thead>
<tr>
<th>Laterality</th>
<th>Number of patients</th>
<th>% out of patients with fibrous dysplasia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unilateral</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>11</td>
<td>91.66</td>
</tr>
<tr>
<td>Bilateral</td>
<td>1</td>
<td>8.33</td>
</tr>
</tbody>
</table>

*Table 7: Distribution of the patients with fibrous dysplasia (n = 12 patients) according to the laterality of the lesion.*

Regarding the maxillary involvement, all the four patients (33.3%) showed buccolingual expansions with cortical thinning and three patients (25%) showed involvement of maxillary antrum in the form of obliteration.

Regarding the mandibular implication, all of the two patients showed involvement of lower border of mandible (16.66%) with involvement of alveolar margin in both cases. All of them showed displacement of overlying teeth (16.66%).

Regarding orbital involvement the two patients showed extraconal extension (16.66%), one of them show involvement of optic nerve (8.33%), and compression of medial rectus muscle (8.33%) as shown in Table 8.

<table>
<thead>
<tr>
<th>Orbit</th>
<th>Number of patients</th>
<th>% out of patients with fibrous dysplasia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Optic nerve</td>
<td>1</td>
<td>8.3</td>
</tr>
<tr>
<td>Extracoanal extension</td>
<td>2</td>
<td>16.6</td>
</tr>
<tr>
<td>Compression of medial rectus</td>
<td>1</td>
<td>8.3</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>33.2</td>
</tr>
</tbody>
</table>

*Table 8: Distribution of patients according to orbital involvement (n = 2 patients).*

Regarding sinuses involvement all of the 4 patients (33.33%) showed obliterated sinuses and none of patients show small or hypoplastic sinuses.

According to the skull base affection, 4 patients showed skull base affection only one patient were presented by isolated skull base fibrous dysplasia whereas 3 patients out of 12 showed skull base affection beside other bony involvement (maxillary, mandibular, petrous, etc). The foramen ovale and foramen rotundum encasement are seen in three patients with less frequent involvement of the other skull base foramina. Middle ear cleft is affected in two patients as shown in Table 9.

---

Table 9: Distribution of the patients with fibrous dysplasia according to involvement of skull base structures (n = 4 patients).

According to the radiological features, all of the patients showed the typical ground glass density with the majority (7 out of 12 patients) shows sclerotic pattern (58.33%) and the rest (5 out of 12 patients) shows mixed lytic and sclerotic pattern (41.66%) as shown in Table 10.

Table 10: Distribution of the patients with fibrous dysplasia (n = 12 patients) according to the radiological features.

Regarding the matrix the majority of patients (7 out of 12 patients) showed homogenous matrix (58.33%) while the rest (5 out of 12 patients) showed heterogenous matrix (41.6%), regarding the margins the majority (8 out of 12 patients) showed well well defined margins (66.6%) while the rest (4 out of 12 patients) showed well defined margins (35%), regarding the cortex the majority (7 out of 12 patients) (58.3%) showed thin cortex, fewer percentage (4 out of 12 patients) showed intact cortex (25%) while only one patient showed absent cortex (8.3%) and no one showed thick cortex as shown in Table 11.

Table 11: Distribution of patients according to radiological features.

Two out of 12 patients are associated with soft tissue swelling (18%).

Figure 13: 19 years old female patient with known fibrous dysplasia complaining of left sided painless cheek swelling figure (a) Axial section showing diffuse expansive mixed lytic and sclerotic ground glass density lesion involving the mandible notably the left side with implication of its alveolar margin and displacement of overlying teeth, (b) Axial section shows the lesion involving the maxillary bone notably the left side with implication of alveolar margin and buccolingual expansion (double arrow heads) with displacement of overlying teeth and implication of the incisive foramen (white arrow) crossing to the right side. (c) Axial section showing the involvement of sphenoid bones along with the nasal vault (cross figure) and frontoparietal regions bilaterally (more left sided) with implication of the orbital bone frame works and skull base of the anterior and middle cranial fossa. The lesion mainly involving the diploic spaces with no cortical interruption noted. Totally encasement of the still patent skull base foramina is seen, namely foramen ovale (black arrow) on both sides, foramen spinosum (curved white arrow) on left side, (d) Axial section showing free foramen spinosum in right side (curved right arrow) bilaterally. Figure (e) Axial section shows encasement of the still preserved superior (striped right arrow) orbital fissures. (g) Axial sections showing encasement of the still preserved optic canals in both sides (notched right arrows in figure f and g). Figure (h) Axial section showing encasement of the still preserved foramen rotundum (bent arrow) and vidian canals (arrow head). Figure (l) Sagittal section showing involvement of maxillary (oval figure), mandibular (cross figure), nasal vault, fronto-parietal bone (black arrows) and sphenoid bone. Figure (j) 3D volume rendering showing distorted facial osseous structures with the previously described expansive lesion of the maxillary, mandibular, nasal and orbital bony walls as well as fronto-parietal bones mainly left side.
Figure 14: 22-year-old female patient complaining of headache and paranasal discharge and forehead swelling (a) axial CT section showing diffuse expansile mixed lytic and sclerotic osseous lesion involving the right nasal cavity with deviation of nasal septum to left side (white arrow). b) axial section showing complete obliteration of the right ethmoidal air cells (bent arrow). c), d) axial sections showing complete obliteration right frontal bone (cross figure) is seen. Distorted outlines of the right orbital bonyframe work with optic canal implication are noted (black arrow). (e) coronal section showing the lesion involving the right inferior turbinate (cross figure), lateral wall of the right nasal cavity with involvement of right orbital floor (notched left arrow) and parietal bone (white arrow). (f) sagittal section showing involvement of the right fronto-ethmoidal (black arrows) regions as well as the right side of the anterior cranial fossa. Clival involvement is seen as well (curved left arrow). (g) 3Dvolume rendering image showing the previously described lesion with right orbital and nasal disfigurement... features suggestive of fibrous dysplasia.
Ossifying fibroma

Regarding sex distribution ossifying fibroma was found more in males with the majority in the age range 10 -< 20 as shown in Table 12.

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>10 - &lt; 20</td>
<td>M: 2 F: 0</td>
<td>2</td>
</tr>
<tr>
<td>20 - &lt; 30</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>30 - &lt; 40</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>3</td>
<td>3</td>
</tr>
</tbody>
</table>

*Table 12: Distribution of patients with ossifying fibroma according to age and sex.*

Regarding the complaint two out of three patients complained of headache, nasal discharge, nasal obstruction and hyposomia (66.66%) with only one patient (33%) complained of proptosis and one patient complained of limitations in ocular mobility as shown in Table 13.

<table>
<thead>
<tr>
<th>Complaint</th>
<th>Number of patients</th>
<th>% out of patients with ossifying fibroma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>2</td>
<td>66.66</td>
</tr>
<tr>
<td>Nasal complaint</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Paranasal discharge</td>
<td>2</td>
<td>66.66</td>
</tr>
<tr>
<td>Nasal obstruction</td>
<td>2</td>
<td>66.66</td>
</tr>
<tr>
<td>Hyposmia</td>
<td>2</td>
<td>66.66</td>
</tr>
<tr>
<td>Orbital complaint</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Proptosis</td>
<td>1</td>
<td>33</td>
</tr>
<tr>
<td>Limitations in ocular mobility</td>
<td>1</td>
<td>33</td>
</tr>
</tbody>
</table>

*Table 13: Distribution of Patients According to Complaint.*

Regarding the laterality of lesions two out of three patients show bilateral involvement (66.66%) and only one patient shows right sided ossifying fibroma.

Regarding the areas involved two out of three patients showed sinuses and nasal cavity involvement whereas only one patient showed orbital and another one showed anterior cranial fossa involvement.

Regarding the sinuses involvement two out of three showed obliterated and retained secretions (66.66%).

Regarding the orbital involvement there were one patient (33%) whom presented with proptosis and another one showed displacement of right medial rectus.

Regarding the radiographic features two out of three patients (66.66%) showed mixed lucent and sclerotic density whereas only one showed cystic density. Regarding the matrix the three of them showed heterogenous matrix. and regarding the margins the three of them showed well defined margin and regarding the cortex 2 patients showed thin cortex whereas one patient showed intact cortex.
**Figure 15:** 15y Male patient with known ossifying fibroma complaining of proptosis and headache. (a) axial section show a large sino-nasal complex soft tissue fibro-osseous lesion epicentered upon the ethmoidal air cells (notably right sided) (cross figure) displaying faint ossific densities and hypodense areas. Laterally, it is violating the medial wall of the right orbit with extraconal component and consequent proptosis (white arrow). (b) coronal section showing superior extension where, it violates the ethmoidal roof with consequent intracranial extension within the anterior cranial fossa (curved white arrow). Inferiorly, it is extending within the right nasal cavity with remodeling of its lateral wall and consequent projection within the right maxillary sinus (black arrow). Medially, it is encroaching upon the left nasal cavity with retained secretions (left white arrow). (c) sagittal section showing Posterior extension where it projects within the sphenoid sinuses (black arrow). Limited extension through right posterior choana is seen (white arrow). Anteriorly, it is protruding into the frontal sinuses. Consequent opacification of the previously mentioned sinuses with retained secretions is seen.
Figure 16: 15 Male patients with ossifying fibroma complained of headache, nasal discharge and obstruction. (a) axial section showing a well-defined fibro-osseous lesion, epicentered upon the right anterior ethmoidal air cells (black arrow). It shows predominately dense sclerotic pattern with scanty lucent areas. (b) coronal section shows Inferior extension of the lesion, where it extends into the right nasal cavity (curved white arrow in b) with posterior displacement of the middle turbinate. Supero-laterally it violates the lamina papracia (black arrow) with extra-conal component displacing the right medial rectus muscle still no definite global compromise seen. (c) coronal section showing inferolateral extension of the lesion where it implicates the right osteo-meatal complex with mild extension into the right maxillary sinus (arrowhead). Intact ethmoidal roof is noted (black arrow). Consequent opacification of the right maxillary sinus (cross figure) with retained secretions are seen.
Fibro-Osseous Lesions of Cranio-Facial Complex

Osteoma

9 patients were diagnosed to have osteoma (39%) in the present study. Regarding the complaint two out of nine patients were presented by mass/swelling (22%), whereas most of them 8 patients (88.88%) were presented by headache and only one patient was presented by recurrent otitis media (11%) as shown in Table 14.

<table>
<thead>
<tr>
<th>Complaint</th>
<th>Number of patients</th>
<th>% out of patients with osteoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mass/swelling</td>
<td>2</td>
<td>22</td>
</tr>
<tr>
<td>headache</td>
<td>8</td>
<td>88.88</td>
</tr>
<tr>
<td>Recurrent OM</td>
<td>1</td>
<td>11</td>
</tr>
</tbody>
</table>

*Table 14: Distribution of patients according to complaint.*

According to the laterality of the lesions, all the diagnosed patients had unilateral lesion with five patients (55%) had right sided lesions in comparison to four (44%) patients with left sided lesions.

Regarding the site of predilection, the frontal sinus (44%) was found to be the most affected portions of the cranio-facial complex with less frequency regarding the other facial regions as shown in Table 15.

<table>
<thead>
<tr>
<th>Site</th>
<th>Number of patients</th>
<th>% out of patients with osteoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mandible</td>
<td>1</td>
<td>11</td>
</tr>
<tr>
<td>Frontal</td>
<td>4</td>
<td>44</td>
</tr>
<tr>
<td>Ethmoidal</td>
<td>1</td>
<td>11</td>
</tr>
<tr>
<td>High parietal</td>
<td>1</td>
<td>11</td>
</tr>
<tr>
<td>External auditory canal</td>
<td>1</td>
<td>11</td>
</tr>
</tbody>
</table>

*Table 15: Distribution of the patient with osteoma according to site of predilection.*

Regarding the radiological features, six patients (66.66%) showed dense sclerotic matrix whereas three patients show mixed lytic and sclerotic (33%). as shown in Table 16.

<table>
<thead>
<tr>
<th>Radiological feature</th>
<th>Number of patients</th>
<th>% out of patients with osteoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dense sclerotic</td>
<td>6</td>
<td>66.66</td>
</tr>
<tr>
<td>Mixed lytic and sclerotic</td>
<td>3</td>
<td>33</td>
</tr>
</tbody>
</table>

*Table 16: Distribution of patients according to radiological feature.*

Regarding the matrix most of patients almost 6 patients (66.66%) showed homogenous matrix and only 3 patients showed heterogeneous matrix (33%) and regarding the margins all patients showed well defined margins (100%) and none showed ill-defined margins (0%).

Regarding degree of sinus involvement, two patient showed osteomas (22%), completely occupying the whole sinus where as one patient showed associated retained secretions (11%) and the rest of sinuses were free.

Regarding the shape of the osteomas, about eight patients (88.88%) were presented with sessile lesions, while one patient (11%) was presented with pedunculated lesion.

**Figure 17:** 33 years old female patient complaining of mandibular osteoma. (a) axial (b) coronal and (C) sagittal CT cuts, (d) 3D VR image showing an ovoid mass lesion (white arrow) arising exophytically from the left angle of the mandible the mass shows homogenous dense osseous attenuation (1500 HU) and associated with sclerosis of the marrow spaces of the mandibular angle (curved arrow). It grows in and expands the caudal part of the masseter with no intra-muscular extensions...
Fibro-Osseous Lesions of Cranio-Facial Complex

Figure 18: 64 years old male patient complaining of frontal osteoma (a) axial (b) coronal (c) sagittal CT sections, (d) 3D VR (a) axial section shows expansile osseous lesion protruding within the right frontal sinus (black arrows). It shows homogenous dense sclerotic matrix. (b) coronal section shows lesion with encroachment upon frontal outflow tract (white arrow) and (c) sagittal section shows retained secretions within frontal sinus (curved arrow). (d) 3D volume rendering shows osseous structure protruding from frontal sinus.

Discussion

Fibro-osseous lesions (FOLs) of the maxillofacial bones comprise a diverse group of pathologic conditions that includes developmental lesions, reactive or dysplastic diseases, and neoplasms [53].

Fibro-Osseous Lesions of Cranio-Facial Complex

It can involve paranasal sinuses, skull base, maxillofacial region. Fibro-osseous lesions involving skull base and paranasal sinuses are uncommon clinical findings. The most common entities within this group are fibrous dysplasia (FD), ossifying fibromas (OFs) and osteomas [54].

The current study was conducted on 25 patients. They were 10 males and 15 females. Their ages ranged between 15 and 60 years with a mean age of 40 - 43 years.

Most of the cases in this study was pathologically proven to be fibro-osseous lesions and surgically operated. The diagnosis is made mainly by combination of radiological features, clinical and pathological features.

A different approach to morphological features in FOLs has been used where the newly developed technique of computer generated three-dimensional (3D) reconstruction to evaluate the shape and quantity of osteoid tissue within the lesions was applied. 3D images are remarkably informative in many areas of clinical practice, although their benefits in FOLs are debatable. Radiographic images of FOLs are complex because of the mixture of fibrous connective tissue and osteoid tissue. Therefore, it is hypothesized that 3D findings could help identify morphological features in these lesions [55].

It has to be underlined that 3D images aren’t superior to 2D images. As the original 2D information is reduced by the post processing, diagnosis should not be made based on the 3D images alone. But as a complementary examination 3D images help to make a precise diagnosis in cases of complex pathologies. Furthermore, 3D images are a helpful tool for presentation of pathologies [56].

In the present study, the cranio-facial fibrous dysplasia that was diagnosed in about twelve patients (48%) and osteoma that was diagnosed in about nine patients (36%) take the upper hand regarding the final diagnosis whereas ossifying fibroma are only three patients (12%) and one case of brown tumour (4%).

Fibrous dysplasia, ossifying fibroma, and osteoma are three distinct entities that lie along a continuum containing the least to the most bony content [3]. The diagnosis of fibrous dysplasia and ossifying fibroma is made by correlation between clinical, radiologic, and pathologic findings.

Fibrous Dysplasia (FD) is a non-neoplastic developmental hamartomatous disease of the bone. With an incidence of 1:4000 - 1:10.000 it seems to be a rare disease with a low number of cases. However, it represents approximately 2.5% of all bone lesions and in total about 7% of all benign bone tumors [57].

The disease has three different subtypes (monostotic, polyostotic, McCune Albright Syndrome).

Craniofacial fibrous dysplasia (CFD) is often found in childhood and usually stabilises in adulthood. In special cases progress of the lesion is possible up to the seventh decade, but is reported to be very uncommon.

The main presentation of patients with CFD is headache as seen in 8 patients out of 12 (66.6%), swelling in the affected region in four patients (33.3%) and facial deformity as seen in only two patients (16.6%).

Craniofacial involvement is found in about 10% of cases of monostotic FD. In this study about 50% of the cases were monostotic and the other 50% were polyostotic. Regarding the site of implication of the cranio-facial fibrous dysplasia, the maxillary bone involvement is seen in four patients out of 12 (33.3%) is most frequently affected, followed by equal distribution for the mandibular (16.6%), petrous (16.6%) and sino-nasal bones (16.6%). The skull base, zygoma and occipital condyle are less frequently affected in the present study (only one patient each 8.3%).
The progression of the lesion may cause aesthetic impairment and deformities and clinical symptoms such as visual disturbances, proptosis, orbital dystopia, nasal malfunction, dental problems and sensory disturbances in the affected regions. FD may radiographically and histologically resemble cherubism, central giant cell granuloma, and other giant cell lesions.[58].

This study agrees with some authors that the craniofacial type of fibrous dysplasia is found to be as common as fibrous dysplasia of the jaw and was more commonly seen in younger age (11,19,22 years) as shown in our study. The unilateral nature of fibrous dysplasia was noted in most of cases (91.6%) only one patient was bilateral (8.3%) [59].

The most commonly involved cranial bones were maxilla and frontal bones. This study also agrees with some authors that when the maxilla was affected, other adjacent bones separated by sutures such as zygomatic, sphenoid, frontal, and nasal bones might also get affected [60]. This study showed that the right side was more commonly involved as shown in 11 patients (91.6%) than the left (only one patient 8.3%), which was similar to the findings of previous studies [2].

Eversole, et al [2] suggested that the ill-defined margins of the lesion helped to differentiate it from other fibro-osseous lesions. In this study, the margins were ill-defined in eight patients out of twelve (66.6%) except in the region where they extended to the cortex of bone.

This study agrees with some authors that ground glass appearance was the most common radiographic appearance of internal structure of fibrous dysplasia which substantiated the diagnosis of fibrous dysplasia [16]. In this study about 7 patients out of twelve (58.3%) shows sclerotic matrix and 5 patients (41.6%) shows mixed lytic and sclerotic and only 2 patients (16.6%) show radiolucency which support the ongoing study at National Institute of Health (NIH) [61] which demonstrated that the typical characteristics of FD on CT and the natural radiographic progression may vary from a “ground-glass” or homogenous appearance to a mixed radio-dense/radio-lucent lesion as the patient ages. In pre-pubertal patients with polyostotic fibrous dysplasia (PFD) or McCune Albright Syndrome (MAS), the lesions most often appear as homogenous, radio-dense lesions on CT. As these patients enter the second decade of life, the FD lesions progress to a mixed appearance, which stabilizes in adulthood but does not resume a homogenous appearance. While the change to a mixed radiographic appearance alone does not require further biopsy or investigation, we recommend careful monitoring and intermittent craniofacial CT during the pubertal phase of the young patient [62].

Fibrous dysplasia shows bucco-lingual expansion in all four patients with maxillary involvement (33.3%) causing thinning of the cortical plate. The expansion of the external surface of the affected bone assumed a grosser but still recognizable anatomical shape. The lesion displaced the inferior alveolar canal in all four directions (buccal, lingual, superior, and inferior) in the four patients with maxillary involvement (33%). The displacement was in contrast with the finding of Petrikowski who suggested that the upward displacement of inferior dental canal was a unique characteristic of fibrous dysplasia. However, the displacement of the canal in various directions might be explained by the location of epicenter in relation to the canal. For example, if the epicenter was superior to the inferior alveolar canal, it would displace the canal inferiorly.

The lesion involving maxilla showed expansion on the external surface and on internal surface into maxillary sinus. The expansion of maxilla reduced the size of maxillary sinus cavity although the shape seemed unaltered. This unique finding could help in differentiating fibrous dysplasia from other tumors such as ossifying fibroma [62]. Fibrous dysplasia might also completely obliterate maxillary sinus and displace the floor of orbit.

This study agrees with some authors that say the sinuses may be affected by FD, with the most frequent site being the sphenoid sinus, followed by the ethmoid and maxillary sinuses [8]. This is not surprising, as the anterior cranial base is often affected in patients with craniofacial PFD [8]. The entire sinus can be completely obliterated by FD, yet surprisingly the incidence of sinusitis is not greater than the general population in these patients. This may be explained by the loss of air space and Schneiderian membrane in an obliterated sinus and the elimination of a source of infection.

The features of fibrous dysplasia such as the margins, internal structure, and effect on surrounding structure were well characterized on CT images. The margins of lesion, ground glass appearance, and displacement of maxillary sinus were characteristic and consistent with the findings of fibrous dysplasia. Although no single radiographic feature is pathognomonic of fibrous dysplasia, all the features are suggestive of fibrous dysplasia.

CT accurately establishes the diagnosis and extent of bone involvement. Involvement of optic canals, orbital fissures, frontonasal ducts and osteomeatal complex can be best evaluated by CT scanning. CT characteristics of fibrous dysplasia include expansion of the involved bone with heterogeneous pattern of CT densities associated with scattered or confluent islands of bone formation. CT attenuation levels have been reported to range from 34 to 513 HU similar to our study depending on the fibrous tissue and bone content [63].

CT imaging is recommended to define the anatomy of individual lesions and to establish the extent of disease. A standard craniofacial CT, without contrast and with slice thickness no greater than 3.75 mm (from top of the head to the thyroid region), is used to evaluate for the presence of FD in the skull base and facial bones. Historically, plain films of the craniofacial region were used but because of the overlapping of adjacent structures, involvement of the skull base was often underreported [15].

The FD lesions of the face may be described as quiescent (stable with no growth), non-aggressive (slow growing), or aggressive (rapid growth +/- pain, paraesthesia, pathologic fracture, malignant transformation, association with a secondary lesion). In the case of a quiescent FD lesion in which the patient does not complain of facial deformity, observation and monitoring for changes is an acceptable treatment modality. Annual evaluations may be adequate. The patient's concerns and symptoms, clinical assessment including sensory nerve testing in the region of involvement, photographs, and facial CT should be obtained at each visit. An annual CT may be necessary for the first 2 years; however, the interval may be lengthened based on the clinical findings [6].

Surgical contouring by a maxillofacial or craniofacial surgeon is indicated if the patient is bothered by facial disfigurement. While complete resection may be possible in monostatic lesions, it is unlikely to be possible in PFD or MAS, and the surgeon must weigh the reconstruction options that will provide the patient with the best outcome as well as preserve the function of adjacent nerves and structures.

These patients may also require orthognathic surgery to correct a concurrent malocclusion or facial/dental canting (slanting or oblique surface) [10]. There is no documented contraindication for orthognathic surgery so long as the lesions are quiescent. Bone healing appears to be normal with conventional rigid fixation. Regular follow-up with the surgeon is necessary to determine that there is no recurrence and further deformity.

In patients with non-aggressive but active fibrous dysplasia (FD), it is ideal to wait until the lesion becomes quiescent and the patient has reached skeletal maturity before performing an operation. However, in cases where the patient's psychosocial development may be impaired due to the facial deformity, surgical contouring and/or resection may be warranted. The patient and family must be aware of the potential for regrowth if the lesion cannot be resected completely, which is often the case. In cases of polyatomic fibrous dysplasia (PFD) or McCune Albright Syndrome (MAS) where the disease is extensive, the lesions are often not respectable. Repeat surgical contouring and extensive debulking may be necessary to achieve acceptable facial proportions [11].

In the future, improvement in CT imaging and software will allow for accurate surgical simulation and intraoperative navigational tools may guide the surgeon throughout the contouring. Advanced CT software is useful for superimposition of pre- and postoperative images. These can then be compared to followup CT scans to determine stability of the result or the presence of regrowth. Despite these new imaging technologies, there is no therapy or technology that can predict and/or prevent regrowth.

Ossifying Fibromas

Ossifying fibromas (OF) of the craniofacial skeleton, as described in WHO classification of odontogenic tumors (2005) [64] are benign fibro-osseous neoplasms characterized by the replacement of normal bone by a fibrous cellular stroma containing foci of mineralized
bone trabeculae and cementum-like material that vary in amount and appearance [53]. The accurate nature and classification of OF has undergone considerable debate among pathologists, resulting in a confusing evolution of competing nomenclatures [53].

According to their pattern of mineralization, four overlapping clinicopathological entities have been historically identified: juvenile psammomatous ossifying fibroma (JPOF), juvenile trabecular ossifying fibroma (JTOF), gigantiform cementoma (GC) and cemento-ossifying fibroma (COF) not otherwise specified (NOS).

The conventional ossifying fibroma usually presents as a solitary, slow growing, monostatic tumor in the third and the fourth decades of life in contrast. It shows female predilection and the male to female ratio is around 1:5 [65], whereas in our study the three cases were male and they were of young age 15y and 32y. Although found predominantly in the mandible (75%), it can also arise in the skull base and PNS [65] as shown in our three cases. It has also been reported to occur in the temporal bone. The tumor is known to be more aggressive in young patients [66]. In the jaws, the tumor shows affinity for the molar area [66]. In the mandible, it can give rise to pathological fractures and osteomyelitis in the long run if left untreated [67].

The juvenile variants of ossifying fibromas share many similarities, but they have been distinguished on the basis of their histopathological features, site, and age of recurrence [68]. Their location is also different: juvenile psammomatoid ossifying fibroma (JPOF) arises mainly around paranasal sinuses as shown in our three cases and orbits, whereas juvenile trabecular ossifying fibroma (JTOF) usually affects the maxilla. The last entity, cement-ossifying fibroma (COF), is an odontogenic neoplasm arising from the periodontal ligament and affects the tooth-bearing areas of the jaws, mandible, and the maxilla; the cementicles are the characteristic feature instead of the bone elements. JTOF also known as trabecular desmo-osteoblastoma affects mainly the jaws of children and adolescents. Only 20% of the patients are over 15 years of age.

This study agrees with [26,28,69] in a review of a number of case series the mean age range was found to be 8.5 - 12 years whereas in our study it was about 15 years. Origin in extra gnathic locations is extremely rare.

Orbital extension of sinonasal tumors may result in proptosis as shown in one of our cases (33%) and visual complaints including blindness, nasal obstruction (66.6%), ptosis, papilledema, and disturbances in ocular mobility (33%). This study agrees with [25] that radiographic examination of JPOF shows a round, well-defined as shown in our three cases, sometimes corticated osteolytic lesion with a cystic appearance as shown in one of our cases (33%) whereas (66%) showed mixed lytic and sclerotic. Sclerotic changes are evident in the lesion which may show a ground-glass appearance. The lesions appear less dense than normal bone. This study disagrees with other studies which stated that the walls of the involved sinuses may undergo further remodelling and thickening, sometimes along with erosions [70] as in two of our cases there were thinning of the cortex (66.6%).

Clinically, it is often characterized by a progressive and sometimes rapid expansion of the affected area; pain is a rare symptom. Cystic degeneration and aneurysmal bone cyst formation has been reported in a few cases. Radiographically, juvenile trabecular ossifying fibroma (JTOF) is an expansive lesion and may be fairly well demarcated, with cortical thinning and perforation. Depending on the amount of calcified tissue produced, the lesion will show varying degrees of radiolucency or radiodensity. Ground-glass as well as a multilocular honeycomb appearance has been described.

Interestingly, some authors have suggested that all the fibro-osseous lesions relating to ossifying fibroma and its subtypes should be referred to as ossifying fibroma only since they claim that there is no difference in behavior between the subtypes and the histological designations are only academic [71].

Even though some authors opine that fibrous dysplasia and ossifying fibroma cannot be distinguished by microscopy due to histological overlap [66] others opine that there are many features like the absence of cementicles, lamellar trabeculae and osteoblasts to distinguish them [72]. The differentiation between the 2 entities is of great importance because of their divergent clinical behavior. Fibrous
dysplasia is often polyostotic whereas ossifying fibroma is usually monostotic [73]. Fibrous dysplasia is usually a self-limiting disease and therefore a complete resection is unnecessary and in most instances impossible [74]. Radiographically ossifying fibromas are classically described as circumscribed unilocular lesions with a surrounding rim of eggshell thin bone giving a ‘punched out’ appearance [75]. Irregular thickening and thinning of the edges of this eggshell has been described as ‘mouth-eaten’ appearance [76]. In contrast, fibrous dysplasia has a ‘ground glass’ appearance with indistinct borders that blend imperceptibly into the adjacent bone without expansion or bone destruction [77]. But again, some authors have also described ‘ground glass’ appearance in juvenile active ossifying fibroma [69]. Hence CT scan with the bone window technique is the most essential investigation since it defines the extent of the tumor and the destruction of the surrounding tissues. The differential diagnosis of ossifying fibroma includes fibrous dysplasia, giant cell tumor, giant cell reparative granuloma, aneurysmal bone cyst, osteoblastoma and osteosarcoma.

The overall prognosis with most types of ossifying fibroma appears to be good. Despite their tendency for local invasion and recurrence, there are no reported instances of metastatic disease with the exception of certain subtypes of ossifying fibro myxoid tumor. The development of aneurysmal bone cyst in psammatoid ossifying fibroma has been reported [78]. Malignant transformation is very rare [65]. Meningitis secondary to invasion into the cranial cavity has been reported and rarely even death may occur [31]. Surgery is the mainstay of treatment for all types of ossifying fibroma. Growth of the facial skeleton is an important consideration in the paediatric age group [79]. Extensive tumors in this age group are more difficult to manage since the tumors here tend to be more invasive, aggressive and have more chances of recurrence. Mandibular tumors can be treated with conservative surgery but aggressive surgery is warranted for midface and paranasal sinuses because of their more aggressive behaviour [65]. Even though small tumors can be removed by simple curettage and enucleation, they are better avoided for the fear of recurrence especially in juvenile ossifying fibroma. One study showed a recurrence rate of 28% after curettage [71].

Osteoma is a benign tumor composed of mature compact bone or cancellous bone. This tumor is essentially restricted to the craniofacial skeleton and rarely, if ever, is diagnosed in other bones [36]. This study agrees with some authors that the mandible and paranasal sinuses are the most commonly affected sites in the maxillofacial region [38]. Clinically, this neoplasm may be silent for years without any symptoms and diagnosed only when it becomes large enough as in two of our cases (mandibular and frontal osteoma) or observed coincidentally during radiological investigations as shown [38] as shown in 8 patients out of 9 (88%) of our cases. Osteoma of the jaws may arise on the surface of the bone, as a polypoid or sessile mass (periosteal osteoma), or it may be located in the medullary bone (endosteal osteoma) [80].

Osteoma is the most common benign tumor of the paranasal sinus. Its incidence is between 0.014% and 0.43% [3] in this study, there were 6 patients out of 9 (66%) were diagnosed as paranasal osteomas. It usually grows slowly. However, it may extend to the surrounding structures and cause severe intracranial or orbital complications [36]. Sayan, et al. [37] and Longo, et al. [81] reported that the most frequently affected paranasal sinus of osteoma was the frontal similar to our study as shown in 4 patients out of 9 (44%), followed by the maxillary, ethmoidal, and sphenoidal sinuses where in our study the ethmoid were the second most common sinus followed by frontal (11%) Though turbinate osteoma is very rare, some cases have been reported in literature [82]. Osteomas arising in the paranasal sinuses may cause such symptoms as sinusitis, headache as shown in 8 patients out of 9 (88%) in our study, or ophthalmologic manifestations [80], however in this study, the frontal sinus osteomas were found incidentally except one which causes frontal swelling (11%). Regarding sinus involvement this study shows that only two patients show obliterated sinuses (22%) and one patient (11%) shows retained secretions.

The pathogenesis of osteomas is unclear. Some investigators considered it a true neoplasm, while others classified it as a developmental anomaly [37]. Osteomas exhibit continuous growth rather than growth cessation at adulthood, and this characteristic is the major feature distinguishing them from other bony exostoses [83], supporting a neoplastic origin. In addition, the possibility of a reactive mechanism, triggered by trauma or infection has also been suggested [84].

Fibro-Osseous Lesions of Cranio-Facial Complex

Osteomas, usually asymptomatic, often remain undetected unless incidentally found on a routine radiographic survey or until they cause facial asymmetry or functional impairment [85].

Radiographically, the presence of an oval, radiopaque, well-circumscribed mass as shown in all our cases attached by a broad base or pedicle to the affected cortical bone as shown in one of our cases (11%) is a hallmark of peripheral osteomas [86], whereas the rest (8 patients out of 9) were sessile (88.8%) and about 6 patients out of 9 (66.6%) showed dense sclerotic matrix whereas only 3 patients (33%) showed mixed lytic and sclerotic. The differential diagnosis includes exostoses and several pathologic processes including inflammatory and neoplastic lesions such as a sclerotic pattern of chronic osteomyelitis, peripheral ossifying fibroma, periosteal osteoblastoma, osteoid osteoma, and parosteal osteosarcoma [87]. Distinguishing between exostosis and osteoma may be difficult. The clinical features are essential in determining the final diagnosis since both the conditions present similar histopathological features.

Exostoses are usually multiple and located in areas of attached gingival mucosa. Solitary exostoses are rare and occur frequently associated with a local trauma or in regions that received gingival or cutaneous graft [39]. In the present cases, none of these features were observed.

On the other hand, central osteomas may cause more difficulty in diagnosis. Fibrous dysplasia, central ossifying fibroma, odontoma, osteoblastoma, chondroma, cementoblastoma, Paget’s disease, and central osteosarcoma should be added to the differential diagnosis of central osteomas [87]. Moreover, pain is an important symptom in 30% of central osteomas [87].

In summary, osteomas occur predominantly in the maxillofacial region, appearing as slow growing and well-circumscribed, lobulated masses. However, maxillary osteomas are extremely rare lesions and located mainly on the alveolar ridge. The ideal treatment is conservative surgical removal and recurrence is uncommon.

Important limitations of the present study must be acknowledged. The series was a retrospective correlation of radiologic data limited by the data available inpatient records. The fibro-osseous lesions were managed by multiple surgeons with differing biases on the diagnostic and management strategy for fibro-osseous lesions. Though the positive predictive value for osteoma and Fibrous dysplasia was high, neither reached statistical significance. Inclusion of a greater number of patients would provide the adequate statistical power to better understand the predictive value of preoperative CT imaging for fibro-osseous lesions. This represents an inherent limitation of the analysis, given the rarity of these lesions. Thus, the results should be viewed with caution. These critical questions may only be answered in a large, prospective, multicenter study that would require the accrual of larger number of fibro-osseous lesions. The present study hopefully serves to provide the essential framework for future prospective studies to guide these diagnostic and management decisions.

Conclusion

Fibro-osseous diseases of the craniofacial bones make up a diverse collection of disorders that include neoplastic and non-neoplastic diseases and hereditary and nonhereditary conditions. Fibro-osseous lesions (FOLs) share many similar histopathologic features. Furthermore, a number of other non-fibro-osseous disease processes that develop within the jawbones exhibit findings that may closely mimic those seen in fibro-osseous lesions (FOLs). Thus, a definitive diagnosis of a fibro-osseous lesions (FOL) requires correlation of the histologic features with the clinical, radiographic, and intraoperative findings. Although sub-classification of BFOLs occasionally can be challenging, a proper diagnosis is essential to ensure adequate and appropriate treatment.

In the future, improvement in CT imaging and software will allow for accurate surgical simulation and intraoperative navigational tools may guide the surgeon throughout the contouring. Advanced CT software is useful for superimposition of pre- and postoperative images. These can then be compared to follow-up CT scans to determine stability of the result or the presence of regrowth. Despite these new imaging technologies, there is no therapy or technology that can predict and/or prevent regrowth.

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