

Pediatrics and Children Orofacial Tumors

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Pediatrics orofacial growth and tumors rare conditions and some of these cases was a pathological entity by itself like Kummoona jaw lymphoma, these type of malignancies are not well addressed in maxillofacial surgery, but maxillofacial surgeon should be familiar with the incidence of these type of pathology from the points of clinical features investigation's and managements. The managements of these cases required expertise and skill and wide knowledge. The majority of these tumors presented as a mass or swelling or growth or cystic swelling. We should differentiate between benign as non-cancerous and malignant cancerous type they are life threatening disease. We should carry biopsy beside all necessary investigations required like CT scan for any suspicious disease before commencing any treatment modalities.

Benign growth can be related in some cases to infection, inflammation and fluid collection or swelling or a neoplasm (Tumor). Ameloblastoma could be cystic or solid and could be quiet benign or very aggressive and locally invasive to surrounding soft tissue and behave similar to basal cell carcinoma more common in the mandible, we reported Ameloblastoma in the maxilla in 2 years female treated by enucleation without recurrence. All these tumors are treatable and on other hand few malignant tumors are life threatening disease.

Benign growth very common among pediatrics and children presented as large swelling or small growth in the mucosa such as epulides (Fibrous or Giant Cell Epulis) or granulomatous condition and polyps (Fibro-epithelial polyp), these cases are not tumors but tumor like conditions due chronic irritation with superimposed infection once these causes eliminated lesions were disappeared if not small surgical excision quiet enough to eradicate these tumor like condition without recurrence.

Other benign growth are rare but it does featured as pedunculated large mass obstructing the oral cavity of new borne baby interfere with breast feeding this type known as congenital epulides but nowadays known as granular cell myoblastoma of infancy and considered as neoplastic process, these cases immediately excised as pedunculated growth other type effect the posterior part of the jaw as sessile growth suspected as hormonal and might disappeared after birth. Ectopic salivary gland tumors an aggressive type was reported by us in infant in the infra orbital region distorting the orbit and the eye ball we did excised this tumor and the area reconstructed by Sikalastic biological rubber silicon material, another tumor of ossifying fibroma of the mandible of 4 years female treated by radical enucleation but recurrence was expected.

Swelling of lymph nodes might occur as enlarged lymph nodes in the neck due to recurrent infection from tonsils, adenoids or decayed teeth or multiple nodes does occurred in cases with infectious mononucleosis caused by Epstein Barr Virus (EBV). Benign lesions of soft tissue swelling such as branchial cleft cyst on the side of neck and thyroglossal cyst attached to the hyoid bone, easily diagnosed by asking the child to bring his tongue forward, immediately the thyroglossal cyst bulge with the hyoid bone other cystic lesion like dermoid cyst may involve the floor of the mouth extended to the submental region. Cystic hygroma is congenital in origin appear as soft swelling in the side of the neck may became large and extensive to involve an area from supraclavicular region up to the parotid region associated with recurrence of infection might press on the larynx causing obstruction of airway ,complete excision required by elective surgery. Cystic lesions of the jaw either odontogenic associated with tooth structures as central lesions, these were treated once its became large by marsupialization to prevent damage to teeth buds in the jaws or by inoculation and non-odontogenic fissure cleft

cyst, these required complete inoculation. These cystic lesions required surgical removal owing to their ability for continuing growth and potential for infection and may interfere with growth of jaws and teeth. Other hamartomas of vascular and lymphatic may noticed in the orofacial region sometimes as combined one as hemangioma-lymphangioma quiet common in babies in the tongue, lips cheek or side of the face, these require a specific treatment either by medication or surgery or both, many of these cases due to recurrence of infection became thrombosed and disappeared sometimes sclerosing agent like STD gives satisfactory results.

Malignant tumors were reported either epithelial or non-epithelial mesenchymal or lymphatic [1], they are in general life threatening disease. we reported Kummoona jaw lymphoma, a clinico-pathological entity and lethal condition duration of illness within 2 - 3 weeks, very aggressive effecting one jaw or both in the molar-premolar region as massive fleshy type, terminating the life by passing through debilitating status with anemia, high fever and high ESR. Etiology of this malignancy was not by EBV (Epstein Barr Virus) but by a virus similar to HSV (Herpes simplex virus) these viruses was detected by electron microscopy, beside serological studies for detection of Early Antigen (EA), Nuclear Antigen(NA) and Epstein Barr Nuclear Antigen (EBNA), imprints cytology was used for quick diagnosis of the jaw lymphoma and can be treated in the early stage by CHOP based on modified author staging [2-4]. Also other types of malignancy observed in the orofacial region like acute myeloid leukemia, fibro sarcoma, myxo-sarcoma, giant cell angio- sarcoma, osteogenic sarcoma, retinoblastoma, embryonic rhabdomyosarcoma, all these tumors are life threatening malignancies and most of these tumors were treated by chemotherapy or by radical surgery with chemotherapy and deep X-ray therapy and the prognosis depend on staging of the tumor, and aggressiveness of the malignancy. Malignant giant cell tumor of the lower jaw is locally aggressive was treated by radical excision of the tumor and immediate reconstruction by rib graft showed good prognosis. Cases with Kummoona jaw lymphoma can be treatable like stage I and Stage II and our therapeutic management was based on NCI recommendation (Vincristine 1.5 mg/2m, Adriamycin 50 mg/2m, cyclophosphamide 1000 mg/2m, methotrexate 10 mg/2m, prednisolone 50 mg/2m) in eight doses, follow-up and duration of therapy for 24 weeks.

Clinical studies with comment

We did review the managements of 60 pediatrics and children tumors of the orofacial region, their age between 2 days to 14 years and they were 30 female and 30 male, 24 cases were benign and 36 cases were malignant, all tumors like condition were excluded from this studies because they are under category of reactive hyperplasia due to chronic irritation and infection. The treatment modalities of benign tumors ranged from complete excision, surgical shaving of bone as in fibrous dysplasia cases and deep X-ray therapy for some benign tumors of 3000rd. In general our plane for managements of malignant tumors were carried out by initial course of chemotherapy for carcinoma (5FU + Toxter + Carboplatin) fallowed by radical surgery and by another 2 courses of chemotherapy, DXT kept as stand by. In pediatrics every tumor required different type of chemotherapy. Another policy applied on pediatric cases, were treated by radical surgery fallowed by DXT without chemotherapy. Reconstruction of the mandible was achieved by rib graft or iliac crest graft or bone graft with re implantation of the condyle that was not involved by tumor. Temporary reconstruction by metal prosthesis was done for cases required chemotherapy or DXT post operatively [2].

There are two type of Rhabdomyosarcoma, adult type affecting the limbs and juvenile type affecting the head and neck region and they are differ in malignancy and prognosis, the embryonic type of infancy, the baby do not survived more than 2 - 3 months. Ewing sarcoma of the jaw quiet rare showing fleshy growth in the mandible, the treatment of these cases by chemotherapy (Vincristine 2 mg/2m, cyclophosphamide 1200 mg/2m, doxorubicin 75 mg/2m or echinomycin 1.25 mg/2m) for nine weeks initially and fallowed by 42 courses of chemotherapy and fallowed by external DXT. Osteogenic sarcoma of lower jaw was treated by chemotherapy and DXT with good result. Ameloblastoma of the maxilla and sinus of baby of 1 year was treated by complete inoculation through cold well luck operation while ameloblastoma of mandible of young child was treated by resection of the jaw and immediate reconstruction by rib or iliac bone graft. Ossifying fibroma of the maxilla and of the mandible were treated by inoculation but there was possibilities of recurrence [5].

Retinoblastoma of the orbit is highly malignant and rare tumors, we reported one case of 4 year child with large tumor involve the whole orbit not only the ball but also the eyelids, it is developmental with hereditary ,there is a strong relation between tumor and dele-

tion of long arm of chromosome (Del 13q 11), we did radical excision of the tumor and the defect was closed by mobilizing the anterior half of temporalis muscle and inserted through window in the lateral wall of the orbit to block inside the orbit and to prevent any communication by blocking optic foramen, the skin was closed by mobilizing two flaps, frontal-orbital Kummoona flap and facial-orbital flap [6].

Giant cell angiosarcoma is highly malignant tumor we reported one case in female of 4 years in the lower jaw involved the whole mandible and floor of the mouth except ascending ramus, the tumor protruded outside simulating the tongue and the tongue pushed back by the tumor to pharynx, the tumor was soft fleshy with high bleeding tendency, radical excision of the mandible from angle in one side to near the angle in other side, the incision used was through reverse T type incision bisecting the midline of the lips [5], eight pints of blood given because high vascularity, a modified K wire used simulating the shape of the mandible for reconstruction and fallowed by DXT, fallow up of the case was for three years surprising the high bleeding tendency during operation but once tumor excised bleeding stop immediately [5]. There are two type of myxosarcoma one central effecting the mandible and treated by radical excision in adult, while in children when the tumor involve the soft tissue in the maxilla required wide excision of soft tissue. There is no specific chemotherapy been applied as standard technique but every malignant tumor required specific protocol to be used? We reported two cases of fibro sarcoma one involve the lateral wall of the orbit in 2 years girl, we did excise the tumor by L shape incision fallow up for 3 years without recurrence while fibro sarcoma of the maxilla extended to the orbit and base of skull in 8 years female, this type is highly malignant with poor prognosis. Impressive advances against pediatric cancer have resulting in growing population of young person and children who are apparently cured of the cancer disease, this statement might be true but our cases with Kummoona jaw lymphoma does not been applied on our cases, the mortality rate was 91.9% and possibilities remain with differences between our Kummoona jaw Lymphoma and Burkett's lymphoma are clinically, etiologically, electron microscopically and managements were different [7].

Current cancer research focused now days on understanding the response and resistance to treatment and apoptosis and cancer treatment depend not only on cellular damages as achieved by chemotherapy and DXT, but on ability of the cell to respond to damages by inducing apoptotic changes in the apoptotic pathway to end with resistance to chemotherapy drugs and radiation. Mitochondria and cell surface receptors mediate the pathway of apoptosis mediated by Bcl2 family protein and the final excursion of cell death is performed by caspase cascade which is triggered by release cytochrome C from mitochondria. Most of the activities in the development of apoptosis drugs were concentrated on apoptosis inducers for treatment of malignancies, one of these drugs Gemzar (gemcitabine), this drug interfere with the growth and spread of cancer cells by inducing apoptosis and an anti-metabolite and also been used with Carboplatin which is specific chemotherapy for treatment of adenocarcinoma of the pancreas [8].



Figure 1a: Retinoblastoma of right orbit of 4 years boy



Figure 1b: Operative procedure by mobilizing Kummoona frontal-orbital flap and facial-orbital flap for closure of skin and anterior part of temporalis muscle transferred for augmentation of the orbital cavity after excision of retinoblastoma tumor .



Figure 1c: One year post-operative.



Figure 2a: A giant cell angio-sarcoma of the mandible and floor of the mouth, of 4 year girl, tumor simulating tongue and the tongue pushed back.



Figure 2b: Specimen of Giant cell angio sarcoma showing extensive tumor involve the whole mandible and floor of the mouth with fleshy highly vascular tissue tumor, a reverse T shape incision used for bisecting the lower lip for radical excision of the tumor.



Figure 2c: Three years post-operative.



Figure 3: Embryonic Rhabdomyo-myosarcoma of baby of 3 months fetal type.



Figure 4a: Kummoona jaw lymphoma involve the right maxilla and orbit of 4 year boy history 3 weeks.



Figure 4b: Three years post chemotherapy of Kummoona Jaw lymphoma.

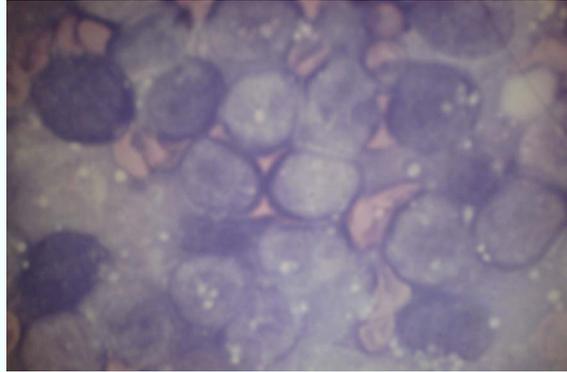


Figure 4c: *Imprint cytopathology of Kummoona Jaw Lymphoma used a quick method for diagnosis by Giemsa stain lymphoblastic lymphoma cells with uniform degree in cell size and cell maturity, darkly stained due to high content of RNA and cytoplasmic vacuoles due to high content of lipid.*



Figure 5: *New borne baby with granular cell myoblastoma (Congenital Epuldes).*

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