Pediatric Giant Invasive Macroprolactinoma Presented as Pituitary Apoplexy; A Case Report and Literature Review

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Abstract

Pituitary apoplexy (PA) is a potentially life-threatening medical emergency and is caused by acute hemorrhage or infarction of the pituitary gland, generally within a pre-existing undiagnosed macro adenoma (especially non-functioning adenoma and prolactinoma); however, it can occur within a normal pituitary or microadenoma. It is a clinical syndrome characterized by headache of sudden and severe onset associated with vomiting, visual impairment and decreased level of consciousness. A high index of clinical suspicion is required for its early diagnosis, timely management of pan-hypopituitarism and prevention of severe neurologic complications.

Keywords: Pituitary Apoplexy; Neoplasms;

Abbreviations

PA: Pituitary Apoplexy; ACTH: Adrenocorticotropic Hormone; TSH: Thyroid-Stimulating Hormone; T4: Thyroxine; FSH: Follicle Stimulating Hormone; LH: Luteinizing Hormone; GnRH: Gonadotrophin-Releasing Hormone; IGF1: Insulin-Like Growth Factor 1; CT: Computerized Tomography; MRI: Magnetic Resonance Imaging; CAB: Cabergoline, DA: Dopamine Agonists.

Introduction

Neoplasms of the pituitary gland are uncommon in prepubertal children and adolescents. They have a reported prevalence of 1:100000 and account for <3% of all intracranial neoplasms [1-4]. At younger than 20 years of age less than 10% (3.5 to 8.5%) of all pituitary tumors are diagnosed with prolactinomas (prolactin secreting tumors) alone accounting for 50-80% of these pituitary tumors [2-4]. These tumors behave differently in children than in adults; in adults one third of pituitary adenomas are functional tumors and they present as microadenomas (<10 mm in size) whereas in children, 95-97% of pituitary adenomas are functional and are quite large at the time of diagnosis [1,5]. Approximately 70% of pediatric pituitary tumors had suprasellar extension at the time of diagnosis and approximately 10% have been characterized as giant tumors measuring > 4 cm [5]. In patients younger than 20 years, approximately 16% pituitary adenomas can develop pituitary apoplexy (PA) [1].

Pituitary apoplexy (PA) is a rare clinical syndrome which is seen predominantly in patients with pituitary adenomas and is caused by acute hemorrhage or infarction of the pituitary gland [1,6-9]. Various presenting symptoms of pituitary apoplexy (PA) are sudden and severe headache (+/-vomiting), visual disturbances, various neurological signs and altered level of consciousness [1,6,7,9,10].

On the other hand, there is an entity known as “subclinical/asymptomatic pituitary apoplexy”. It is an incidental finding of hemorrhage and infarction/necrosis in a pituitary tumor on routine neuroimaging and histopathological examination. It is more common than the acute apoplexy and is seen in approximately 25% of patients with pituitary tumors. It is not a critical condition and its treatment depends on the size and hormonal status (functioning or nonfunctioning) of the pituitary adenoma.

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Case Report/History

Our patient is a 12-year-old girl high school student, who presented to our hospital's emergency department with a complain of mild frontal headache, visual disturbance (left > right) and syncope. Further detailed history revealed that she was having these symptoms 8 months prior to this presentation (1-2 episodes per month) and these symptoms were getting worse over the last two weeks. She was prepubertal at the time. There was no history of galactorrhea, fever, vomiting, seizures or any neurological deficit. She was vitally stable with BP=105/54 mm Hg, HR=86/minute, RR=24/minute, Height=150 cm (25th-50th percentile), Weight=45 kg (50th-75th percentile), BMI=20 (kg/m²), BSA=1.37 m². Her GCS was 15/15. She had pale left optic disc, relative afferent pupillary defect in left eye and decreased left eye visual acuity measuring 20/100 (normal=20/20) on ophthalmological examination. No restriction of extra-ocular muscle movement was noted.

Plain CT brain was done in the emergency department which showed a large predominantly solid, noncalcified hyperdense mass in the pituitary sella and suprasellar region. Some cystic/necrotic components were seen in its central portion. Based on these CT scan findings and age of the patient, this mass lesion was interpreted as a craniopharyngioma. Contrast enhanced MRI brain was performed the next day which showed a large sellar/suprasellar mass (> 4 cm) with haemorrhage of different ages in its central portion. It was having significant mass effect over the optic chiasma and invasion into the right cavernous sinus. Normal pituitary gland was not separately identified. Based on these MRI features, diagnosis of a giant invasive pituitary macroadenoma with internal haemorrhage was made. Possibility of craniopharyngioma was ruled out. Laboratory investigations showed markedly elevated serum prolactin level measuring 43202 mIU/L (normal=109-557). She also had pan hypopituitarism [cortisol=<28 nmol/L (101-536), ACTH=24 pg/ml (≤46), free T4=8.6 pmol/L (9.0-19.0), LH=<0.07 IU/L (1.80-11.78), FSH=0.13 IU/L (3.03-8.08), estradiol=<37 pmol/L (77-921) and growth hormone=0.14 pg/ml (≤13.40)]. Urine osmolality was 179 mOsm/Kg (300-900). This endocrine profile (in combination with MRI features) was interpreted as hyper functional macroprolactinoma with secondary pan hypopituitarism related to tumor mass effect; however, based on combination of imaging features, endocrinology profile and acute clinical presentation of the patient, finally a diagnosis of acute pituitary apoplexy was made.

Later on she was evaluated by a pediatric multidisciplinary team (comprising endocrinologist, neurosurgeon and ophthalmologist). Steroids (hydrocortisone 5 mg, orally twice a day) were started. After 48 hours of steroid initiation, levothyroxine (50 mcg once daily orally) was added. Cabergoline (dopamine receptor agonist) 0.25 mg once a week was also started. After three days, prolactin level was repeated which showed 50% reduction in its level (from 43202 to 21879 mIU/L). Based on this dramatic reduction in prolactin level and stable condition of the patient, multidisciplinary team decided to withhold any surgical intervention.

She continued her medical treatment with laboratory (particularly endocrinology profile) and imaging (MRI brain) follow up at regular intervals, which showed progressive improvement in her prolactin levels and reduction in the size of pituitary lesion. She had normal prolactin level, persistent pan hypopituitarism and a small residual pituitary fossa lesion on her last follow up at approximately 3 years interval from her initial presentation. Follow up ophthalmological evaluations revealed persistent pale left optic disc; however, an improvement was noted in left eye visual acuity from 20/100 to 20/40. After 3 years of her treatment, Premarin 0.625 mg daily (conjugated estrogen) was added in her medication due to lack of menarche. After 3 months of its use, she had menarche and ultimately, Premarin was replaced by Gynera (ethinyl estradiol 0.03mg / Gestodene 0.075mg). Currently she is on Cabergoline (1.5 mg twice a week), Levothyroxine (100 mcg once daily), Hydrocortisone (5 mg a.m. and 2.5 mg p.m (8.87 mg/m²) and (Ethinyl estradiol 0.03mg / Gestodene 0.075mg) once daily for 21 days, then to discontinue for 7 days and after that repeat the same cycle. Our plan is to continue this medical management and follow up with MRI on a yearly basis for the next two years and then every two years if she remains stable and has no new neuro-ophthalmological issues.

Discussion

Two main types of tumors which occur in pituitary fossa of children are craniopharyngiomas (CPs) and pituitary adenomas. Craniopharyngiomas (about 10-15% of all childhood brain tumors) are benign tumors that account for approximately 80-90% of neoplasms found in pituitary fossa of children [2,5]. Pituitary adenomas are slow growing benign neoplasms arising from the anterior pituitary gland and are classified on the basis of their function and microscopic appearance [5,10]. As stated earlier, prolactinoma is the most common...
Table 1: Serum Prolactin Levels (Reference value: - 109-557 mIU/L).

<table>
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<td>714.2</td>
</tr>
<tr>
<td>24/6/18</td>
<td>501.9</td>
</tr>
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</table>

Figure A: Plain CT Brain. A (axial), B (sagittal) and C (coronal) images. Large solid sellar/suprasellar mass with central cystic component. It measures approximately 3.8 x 3.3 x 5.3 cm and is compressing the right side of 3rd ventricle and foramen of Monro causing mild dilatation of right lateral ventricle. It is likely a craniopharyngioma and needs further assessment by an MRI brain.

Figure B: Baseline Contrast Enhanced MRI brain. A (axial T2), B (coronal), C (sagittal T1). D, E and F (Post contrast axial, coronal and sagittal images respectively). Lobulated sellar and suprasellar mass, isointense to grey matter on both T1 and T2 weighted images and showing heterogeneous enhancement. It measures 3.5 x 3.3 x 5.8 cm, extending laterally in to the right cavernous sinus and has significant mass effect over the optic chiasma, floor of the third ventricle and right foramen of Monro resulting in active hydrocephalus. It has central cystic component with fluid-fluid level on both T1 and T2 weighted images which represents blood of different ages.
pituitary adenoma in prepubertal children which is further classified into; microprolactinoma (<10 mm in maximum dimensions), macroprolactinoma (>10 mm) and giant prolactinoma (>4 cm in size or >2 cm suprasellar extension) [3,4,11]. Giant prolactinomas are associated with significantly elevated serum prolactin (PRL) level, usually >1000 ng/ml and occasionally exceeding 40 000 ng/ml, like our case [3,4].

In 1898, Bailey was the first one to describe a case of pituitary tumor associated hemorrhage and in 1950, Brougham, et al introduced the term “pituitary apoplexy” [6,8,9]. First adolescent with pituitary apoplexy was described by Dawson and Kothandaram in 1972 and a case series of nine adolescents with pituitary apoplexy was described by Jankowski, et al in 2015 [1,12].

PA can occur in both functioning and nonfunctioning adenomas, though more frequently seen in nonfunctioning adenomas [1,6,13]. As nonfunctioning adenomas are usually larger than the functioning adenomas, PA is more commonly seen in large nonfunctioning macroadenomas [1,6,8,9]. Prolactinomas, are also susceptible to apoplexy [8]. Cavernous sinus invasion (usually unilateral) seen in 20% of macroadenomas is another important prognostic factor associated with pituitary apoplexy [8,11,14].

Chances of hemorrhage within a pituitary adenoma are five times higher than other intracranial neoplasms [9,10]. Approximately in 50-80% cases, pituitary apoplexy is often the first presentation and patients are unaware of underlying pituitary adenoma [7-9] Although PA is labeled in non-adenomatous lesions it usually occurs in adenomas [8,9] like hypophysitis [15], craniopharyngioma, Rathke’s cleft cyst, sellar tuberculosis [16], metastasis to pituitary (especially from renal cell carcinoma) [17] or even in a normal pituitary gland during the delivery and puerperium [18].

Although pituitary apoplexy generally occurs spontaneously; different conditions including head trauma, drugs (dopamine receptor agonists, estrogen, anticoagulants), surgical procedures (cardiac or orthopedic surgery), angiographic procedures (especially cerebral angiography), dynamic hormone stimulation testing to investigate pituitary function in pituitary tumor patients, pituitary radiosurgery, radiotherapy, hypertension, transient increase in the intracranial pressure, bleeding disorders, pregnancy, sickle cell anemia, lymphocytic leukemia and diabetes mellitus have been reported as predisposing or precipitating factors for pituitary apoplexy in 10-40% cases, usually in the adult population [1,6-9,11,13]. Pituitary apoplexy review by Briet., et al in 2015 showed “dopamine receptor agonists” and “dynamic pituitary hormone stimulation testing” as two most important triggering factors for pituitary apoplexy in patients under 20 years of age [6]. Jankowski, et al [12] identified predisposing factors in 4 adolescent patients (dopamine agonist in three patients and motor vehicle accident in one patient).
Pathogenesis of pituitary apoplexy is not fully understood; however, different intrinsic features of pituitary adenomas (e.g. relatively higher metabolism, limited angiogenesis and blood flow, decreased vascular density, vascular fragility and high intratumoral and intrasellar pressure) make them prone to bleed and undergo infarction [1,9].

**Figure 1:** Pathophysiology of pituitary apoplexy [9].

**Figure 2:** Mechanisms contributing to the clinical picture in pituitary apoplexy [8].

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Clinical presentation depends on tumor size, timing and extent of hemorrhage and pituitary hormonal irregularities. Classically symptoms evolve in a few hours to two days and mostly patients present with sudden severe headache (+/-nausea/vomiting), visual impairment, altered conscious level and cranial nerve palsies [1,6-9,11].

Diagnosis of pituitary apoplexy can be challenging and needs a prodigious level of clinical suspicion to make a clinical conclusion as majority of these patients are unaware of underlying pituitary tumor. Furthermore, above mentioned symptoms can mimic other common neurological emergencies such as subarachnoid hemorrhage, bacterial meningitis, cerebral ischemia, cavernous sinus thrombosis, migraine and hemorrhagic infarction in a Rathke’s cyst [8,9].

Because of rarity of pituitary adenomas in pediatric population, they are usually not taken into consideration in the differential diagnosis of sellar and suprasellar lesions in this age group, which can often mislead to a diagnosis of craniopharyngioma or low-grade glioma, which are primarily treated by surgery, chemotherapy and/or radiotherapy [3]. Just like the case of Gan H.W., et al [3], our case also emphasizes the importance of a routine pituitary endocrine evaluation related to the sellar and suprasellar lesions and this laboratory work up not only helps in documentation of potential tumor-related pituitary dysfunction but also identifies functioning pituitary adenomas.

Nearly 80% of adult pituitary apoplexy patients show one or more anterior pituitary hormone deficiencies at the time of presentation: central hypoadrenalism/hypocortisolism (ACTH deficiency) occurs in up to 70%, TSH deficiency occurs in 50% and gonadotrophin deficiency occurs in 75% of cases [6,8,9,16]. Hyponatremia, which may be secondary to hypocortisolism or inappropriate antidiuretic hormone secretion, is seen in up to 40% of cases [8,9]. Diabetes insipidus is transient and is seen in less than 5% pituitary apoplexy cases [1,8,9]. The most common (however rarely replaced) pituitary deficit after apoplexy is growth hormone deficiency which is seen in almost all patients [19]. Although no exact data is obtainable for determining the likelihood of such hormonal irregularities in children and adolescents with PA; corticotrophic deficiency seems to be a common finding in this age group as reflected by steroid replacement therapy in 43% of reported cases [1].

Impairment of the pituitary function can be caused by the hemorrhagic destruction of the gland which frequently leads to a permanent hypopituitarism; another possibility of pituitary impairment is compression of the residual normal tissue which is usually not permanent and can be recovered by debulking surgery of the pituitary mass. However, some studies have shown that pituitary deficiencies due to pituitary apoplexy, once established, are permanent and have no relation to the management modalities [8,9]. Our case had pan hypopituitarism at the time of presentation which was permanent and required hormone replacement therapy for all except growth hormone.

Computerized tomography (CT) brain is usually the initial imaging investigation in an acute clinical setting because of its easy and wide availability. CT can show a sellar mass in up to 80% cases; however, it is diagnostic in only 21–28% of cases [19]. Magnetic resonance imaging (MRI) is the imaging modality of choice because of its better anatomical details and portrayal of different ages of bleeding and had been found to confirm the diagnosis of pituitary apoplexy in over 90% of the patients [7,10,19]. In a retrospective series, MRI is superior to CT in that, pituitary tumor in 100% and hemorrhage in 88% of cases were recognized and pituitary tumor in 93% and hemorrhage in 21% of cases were recognized in each modality [8,20].

For a more constant clinical understanding of PA and better judgement of different management routes, Pituitary Apoplexy Score (PAS), was proposed by UK Pituitary Apoplexy Guidelines Development Group in 2010 [19]. This scoring system ranges from 0 to 10 and is founded on the level of consciousness, visual acuity, visual field defects and ocular palsies as shown in the table 2. Surgery is indicated in all patients with PAS ≥ 4 [8,9,19].

In 2014, Jho., et al [21] proposed Pituitary Apoplexy Grading System (table 3): grade 1 patients are asymptomatic (subclinical apoplexy); grade 2 patients present with endocrine dysfunction; grade 3 patients present with headache; grade 4 patients present with ocular paresis, while grade 5 subjects present with acute visual defects or altered mental status/low Glasgow Coma Scale, precluding visual assessment. According to this grading system endocrine dysfunction recovers more frequently in patients with grades 1–3, whereas early surgery is recommended in patients with higher grading [8,9,21].

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Variable | Points
--- | ---
**Level of Consciousness**
Glasgow coma scale 15 | 0
Glasgow coma scale <8-14 | 2
Glasgow coma scale <8 | 4
**Visual acuity**
Normal* 6/6 | 0
Reduced — unilateral | 1
Bilateral | 2
**Visual field defects**
Normal | 0
Unilateral defect | 1
Bilateral defect | 2
**Ocular paresis**
Absent | 0
Present — Unilateral | 1
Bilateral | 2
No change from premorbid visual acuity.

*Table 2: Pituitary Apoplexy Score (PAS) [19].*

<table>
<thead>
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<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>No symptoms</td>
</tr>
<tr>
<td>2</td>
<td>Symptoms caused by endocrinopathy</td>
</tr>
<tr>
<td>3</td>
<td>Headache</td>
</tr>
<tr>
<td>4</td>
<td>Ocular paresis</td>
</tr>
<tr>
<td>5</td>
<td>Visual acuity or field deficit (or low GCS score not allowing testing)</td>
</tr>
</tbody>
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*Table 3: Pituitary Apoplexy Grading System [21].*

**Management**

**Management of pituitary apoplexy**

Pituitary apoplexy is a well-known, rare clinical syndrome in children that can be fatal due to adrenal insufficiency; however, it has excellent outcomes, if managed properly [7,10,22]. In the past it was almost considered a neurosurgical emergency globally and traditionally majority of these patients were treated with early surgery [6,22]. Early surgery (3-10 days) is the mainstay of treatment; however, selected patients can benefit from planned/delayed surgery or even from conservative/medical treatment only [22]. Majority of the patients improve with both surgical and conservative management and its optimal management in the acute phase is controversial [7,8,19,23]. After analysis of outcomes of case series of different treatment options of PA in adults, Singh., et al [22] also concluded that most patients have excellent outcome and statistically there is no significant difference between these two therapeutic strategies [1,22]. However, in pediatric and adolescent patients with PA, outcome of these different management options has not been compared on a larger scale and most of the information available in the literature is in the form of isolated case reports or small case series. In literature, there are about twenty cases of pituitary apoplexy in the pediatric age group (below 15 years of age) [10].
Patient should be evaluated carefully by a pediatric multidisciplinary team (including neurosurgeon, endocrinologist and ophthalmologist) before making a management decision (conservative/medical or surgical) [7,8,19]. Approximately 70% of the patients with PA have ACTH deficiency/acute secondary adrenal insufficiency which is the most important cause of mortality associated with PA [19]. Initially, PA patients should be stabilized medically with corticosteroid replacement therapy if needed. In addition to steroid replacement therapy, patients should also be carefully assessed for any fluid and electrolyte imbalance and endocrine deficiencies which should be acutely managed to ensure haemodynamic stability [1,8,11,19]. According to recent retrospective studies, mild and stable visual and neurological signs of PA can improve spontaneously in majority of the patients with conservative management [19]. However, these patients need close careful monitoring and surgical decompression must be considered if these signs do not improve or show any clinical deterioration [19].

Patients with markedly impaired vision (visual acuity and visual field defects) and level of consciousness are candidates for surgery, preferably by an experienced pituitary surgeon (doing five or more transsphenoidal pituitary surgeries/year) within 3-7 days of onset of symptoms [19,22]. Clinically stable patients and patients with failed conservative management are also candidates for this semi-elective transsphenoidal surgery [19]. Early surgery has significantly greater improvement in visual impairment (visual acuity and visual field defects) but not ocular paresis [8,19].

Ocular paresis (III, IV or VI cranial nerves in cavernous sinus) without any visual field defect or reduced visual acuity is not in itself an indication for immediate surgery. Resolution will typically occur within days or weeks with conservative management [19].

As mentioned earlier, approximately 80% of pituitary apoplexy patients have anterior pituitary hormone deficiencies which are usually permanent and have no relation to the management modalities. 75% patients need gonadotropins, 70% patients need corticosteroids and 50% patients need thyroid hormone replacement therapy on a long term basis [6,8,9,16,19]. Regrowth of pituitary tumor and recurrence of pituitary apoplexy are rare and can occur in both conservatively and surgically treated patients [8,9,19,20]. Therefore, all these patients need periodic endocrinological and imaging follow-up to detect any recurrence. Recommended imaging follow up is MRI at 3–6 months after apoplexy, annually for the next 5 years and then biennially [8,9,19].

Management of prolactinoma

Due to their excellent efficacy, dopamine receptor agonists (bromocriptine, cabergoline, pergolide and quinagolide) are drugs of choice as first-line treatment, for all prolactinomas (micro, macro as well as giant adenomas). Among this group, cabergoline (CAB) in particular has less frequent dosing, better-tolerance and higher therapeutic efficiency. Because of these features, it is also considered as initial drug of choice in children and adolescents [3,4]. Surgery (preferably transsphenoidal approach) is currently considered as second-line treatment option after medical treatment with dopamine agonists (DA). The main indications of surgery are drug resistance/intolerance, cerebrospinal fistulas secondary to tumor reduction after medical therapy, rapid loss of vision or cranial nerve paralysis due to intratumoral hemorrhage or pituitary apoplexy [4] Because of excellent response of prolactinomas to DA; currently, radiotherapy has a limited role in their management. A few indications of external radiotherapy are poor response to medical/surgical treatment, contraindication of surgery and malignant tumors [4]. Finally, it must be kept in mind that reduction in tumor size induced by DA may be accompanied by serious complications such as cerebrospinal fluid rhinorrhea, intratumoral hemorrhage/pituitary apoplexy, herniation of brain and +/- optic chiasma into the sella turcica associated with seizures and/or visual alterations and, finally, tension pneumocephalus [4].

Resistance to dopamine agonists can be defined with respect to failure to normalize PRL levels and failure to decrease tumor size by ≥50%. Using these definitions, failure to normalize PRL levels is seen in 24% of those treated with bromocriptine, 13% of those treated with pergolide and 11% of those treated with cabergoline. Failure to achieve at least a 50% reduction in tumor size occurs in about one-third of those treated with bromocriptine and 10–15% of those treated with pergolide or cabergoline. Studies of in vitro cell preparations show that the D2 receptors of resistant tumors are decreased in number but have normal affinity. (Molitch M.E. Dopamine Resistance of Prolactinomas Pituitary 6: 19–27, 2003) [24].
Conclusion

Pituitary apoplexy is a rare serious medical emergency with a highly variable and unpredictable outcome, which is often misdiagnosed at initial presentation. Despite all the advances in diagnostic techniques and treatment options, its timely diagnosis and optimal
management remains challenging due to lack of specific guidelines (particularly for pediatric and young adolescent population), limited individual experience and variable clinical course of the condition. Such patients should be evaluated and managed by a multidisciplinary team, involving endocrinologists, neurosurgeons, neuroophthalmologists and neuroradiologists, to minimize morbidity and mortality. Pituitary endocrine evaluation should be done as a routine work up in all patients with sellar and suprasellar lesions. Majority of PA patients show one or more anterior pituitary hormone deficiencies at the time of presentation and such deficiencies, once established, are permanent and need replacement therapy.

ACTH deficiency/acute secondary adrenal insufficiency which is the most important cause of mortality associated with PA must be promptly treated with stress doses of hydrocortisone. Patients without severe neuro-ophthalmological deficits or with a quick response to the acute management, can be treated conservatively; however, such patients need close careful monitoring and surgical decompression must be considered if these signs do not improve or show any clinical deterioration.

Regrowth of pituitary tumor and recurrence of pituitary apoplexy are rare and can occur in both conservatively and surgically treated patients. Therefore, all these patients need periodic endocrinological and imaging follow-up to detect any recurrence.

Most patients have excellent outcome and statistically there is no significant difference between surgical and conservative management. However, in pediatric and adolescent patients with PA, outcome of these different management options had not been compared on a larger scale and most of the information available in the literature is in the form of isolated case reports or small case series.

Bibliography


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