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RESEARCH ARTICLE

PSAMMOMATOID JUVENILE OSSIFYING FIBROMA OF MAXILLA WITH SECONDARY ANEURYSMAL BONE CYST: A CASE REPORT WITH REVIEW OF LITERATURE.

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Abstract

Psammomatoid juvenile ossifying fibroma is a rare, benign bone-forming neoplasm with tendency to aggressive local growth that is distinguished from other fibro-osseous lesions primarily by its early age of onset, clinical presentation, aggressive behavior and recurrence tendency. The pathognomonic histopathologic feature is the presence of spherical ossicles, which are similar to psammoma bodies. Very few cases occur in association with the secondary aneurysmal bone cyst (ABC) formation have been reported in the literature, we present here an intraoral case in maxilla with secondary ABC in a four years old girl.

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Introduction:-

Maxillofacial fibro-osseous lesions are a group of jaw disorders characterized by replacement of bone by a connective tissue matrix, which displays varying degrees of mineralization in the form of calcifications, woven bone or cementum-like structures indistinguishable from “cementicles” (8). This group includes a broad group of several entities such as ossifying fibroma, juvenile ossifying fibroma, fibrous dysplasia, cement- ossifying fibroma and familial gigantiform cementoma. (22) In general, fibro-osseous lesions of the maxillofacial region present a diagnostic dilemma for the clinician and pathologist owing to their overlapping clinical and histopathological features.

Psammomatoid juvenile ossifying fibroma (PJOF) is an aggressive variant of ossifying fibroma predominating in the pediatric age group with its peak incidence in the first and second decades of life with almost no sex predilection (4). PJOF is a rare entity. In an extensive review by El Mofty in 2002, he stated that 230 cases were reported till then (24). The term psammomatoid ossifying fibroma was originally used by Gogl in 1949 to describe two cases, one affecting the frontal sinus in a 5-year-old boy and the other was in the ethmoid sinus in a 9-year-old girl (21-24). This was done after Benjamins in 1938 who reported a lesion of the frontal sinus, which he termed “osteoid fibroma with atypical calcification” that was histologically similar to Gogl’s cases. Also earlier references by Moser in 1899, Fetissof in 1929, and Ringertz in 1938, had described similar pathological characteristics, but assigned different nomenclature (2). Moser termed the lesion a sarcoma while Fetissof called it “spongy osteoma,” and Ringertz called it “juvenile basal fibroma”. Makeke in 1983 reviewed 86 cases and considered the lesion to be a variant of osteoblastoma and termed it psammous desmo-osteoblastoma and he refered to the trabecular variant as trabecular desmo- osteoblastoma (1, 24). Johnson et al in 1952 was the first to use the term juvenile active ossifying fibroma (3).

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Histiogenesis:-
It has been proposed that the mesenchymal cells of the periodontal membrane serve as multipotential precursor cells capable of differentiation into cementum, osteoid or fibrous tissue and give rise to a spectrum of fibro-osseous lesions (6, 10). On the other hand, according to some investigators, PJOF perhaps originate from improper development of the tissue generating the bony septa between the roots of molar teeth (4). Moreover, Johnson et al hypothesized that the lesion arise from the overproduction of myxofibrous cellular stroma normally involved in the development of septa in the paranasal sinuses as they enlarge and pneumatize (16).

Several studies demonstrated the presence of nonrandom chromosome break points at Xq26 and 2q33 resulting in (X; 2) translocation (3, 4).Recently, Pimenta et al 2006 described haploinsufficiency of new tumor suppressor gene CDC73 (HRPT2) in several cases of PJOF(17).

Clinically:-
Johnson et al reviewed 3000 fibro-osseous lesions and found that 112 met the criteria for the juvenile active ossifying fibroma (6). Among the facial tumors, 90% arise from the paranasal sinuses and the remaining 10% involve the mandible (4). Few cases involving the temporal bone have also been reported. Faizan et al reported that the maxillary tumors often fill and obliterate the maxillary sinus whereas mandibular tumors usually involve the ramus and angle (5). Very rarely has it been reported extracranially (3%).

Maxillary PJOF presents clinically with proptosis, visual disturbances, blindness from compression of the optic nerves and orbit, disturbances in ocular mobility, recurrent headaches, and nasal obstruction.(7) Aggressive growth occurs in some but not all cases encroaching adjacent orbital, nasal, and cranial compartments, distorting the face, displacing orbital contents, and blocking normal sinus drainage to form mucoceles. Intracranial extension develops slowly with few neurological signs or symptoms, even though the lesions may be large enough to fill the anterior cranial fossa (12). The dura maintains an effective barrier to invasion of brain. Occasionally, patients will present with meningitis caused by communication between paranasal sinuses and subarachnoid space.(12) Such behavior may be related to younger patient age and the concurrent development of aneurysmal bone cysts, which is seen in some cases(4).

The radiographic appearance manifests as well-demarcated, unilocular or less commonly multilocular radiolucencies with a variable amount of radiopacity, usually manifesting as fine flakes or as ground- glass opacification (5-14). Aggressive forms tend to show more expansile growth pattern than the typical variety. However, they still tend to exhibit sharp demarcation from the normal adjacent bone and this occasionally reflected by an abrupt loss of the overlying cortex (26). The computed tomography has added value to the diagnosis and is used to confirm the extent of the destruction (18).

On gross inspection, PJOF appears as a yellow-white, homogeneous, and firm lobulated mass with gritty consistency. Cystic spaces, although rare, can be present. Histologically the most characteristic feature is the presence of numerous small, round ossicles or “psammomatoid” bodies that are embedded in a cellular fibrous stroma. (14, 16, 20), the word “psammos’’ is derived from the Greek word meaning “sand” (25). The ossicles are mineralized collagenous foci that vary from small, smoothly contoured round-to-oval patterns to larger, irregular ones (13, 14, 16, 19). A prominent marginal osteoid rim surrounds the ossicles (4).The ossicles are identified within the bony trabeculae as well as within the adjacent cellular stroma. Their number varies from only a few to a dense population of innumerable spherical bodies. Each one of these bodies shows basophilic center with esinophilc periphery with brush borders.

An aneurysmal bone cyst (ABC) can occur as a secondary change in association with a number of benign and malignant bone lesions, the exact number of PJOF cases converting into ABC have not been documented in the literature so far, but Makek in his study found out that of the 69 cases of PJOF, only three cases showed ABC transformation. Based on this study the estimated percentage would be 4.3% (9). Here we report an additional case of PJOF that occurred in the maxilla, and was associated with ABC, which is a rare entity.

Case presentation:-
Four years old girl was examined in the outpatient clinic of oral and maxillofacial department at The Memorial Souad Kafafi MUST University Hospital complaining from unilateral left maxillary swelling that caused maxillary expansion, lateral globe displacement and proptosis fig (1), she also complained from nasal obstruction. Swelling

started from three months and grown rapidly. Past medical and family history was noncontributory. Radiographically the CT showed destructive lesion involving maxilla, maxillary sinus up to the base of the orbit fig (2).

Surgical excision of the tumor was decided under general anesthesia. Through Weber-Ferguson incision the lesion was approached extraorally, hemimaxillectomy was performed that extended to palatal midline and posteriorly to posterior palatal margin fig (3). Complete blunt and surgical dissection was performed till complete excision of the tumor as one piece. Surgical closure was performed with suitable techniques.

Simple acrylic obturator was manufactured to obturate the maxillary defect. A future reconstruction plan is prepared using temporalis muscle flap at the age of growth cessation with titanium mesh.

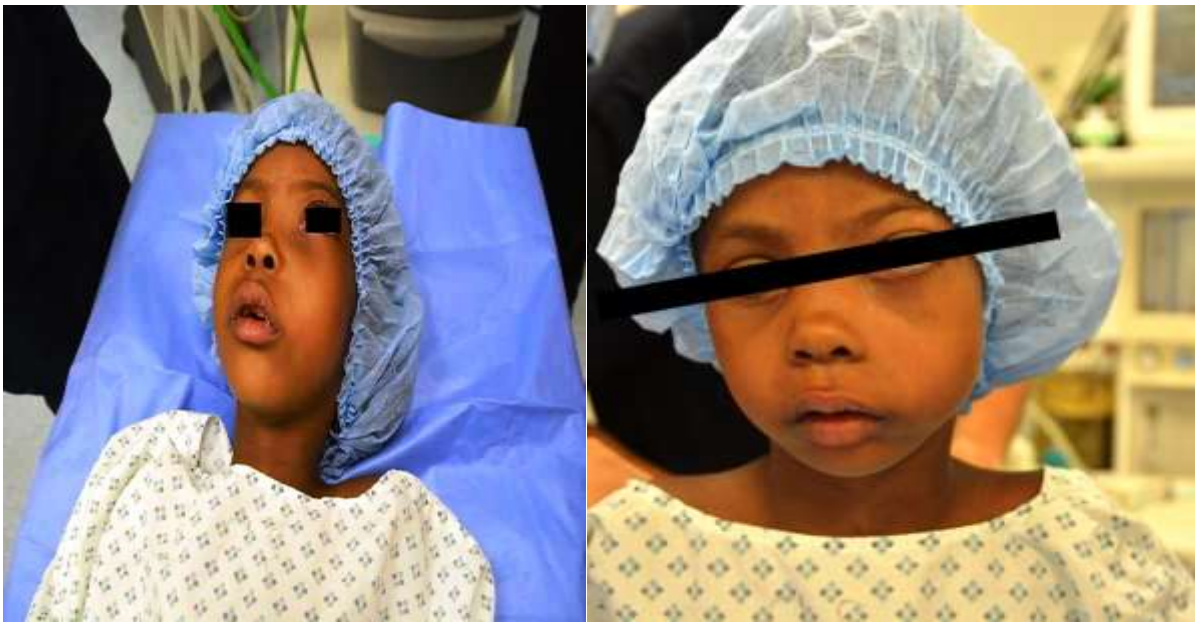

Fig (1):- tumor caused unilateral facial swelling with proptosis.

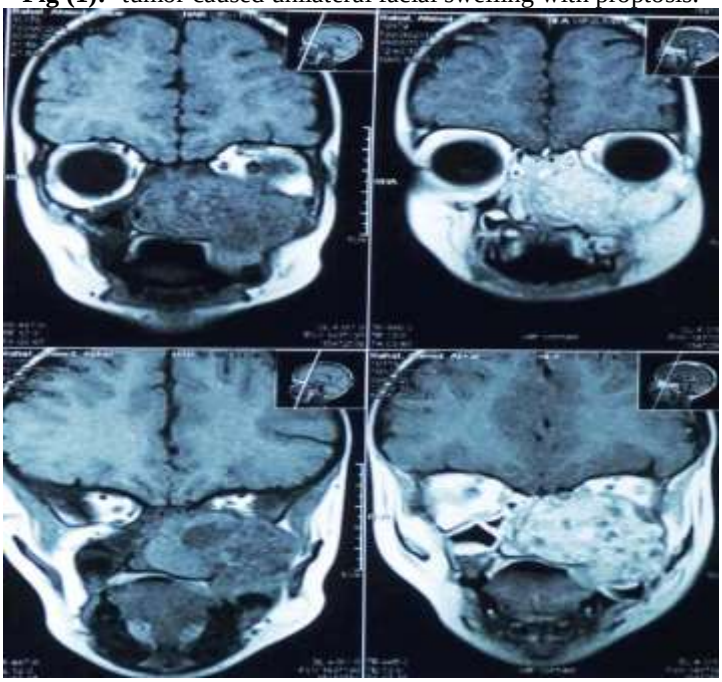

Fig (2):- CT showed destructive lesion involving maxilla, maxillary sinus up to the base of the orbit.

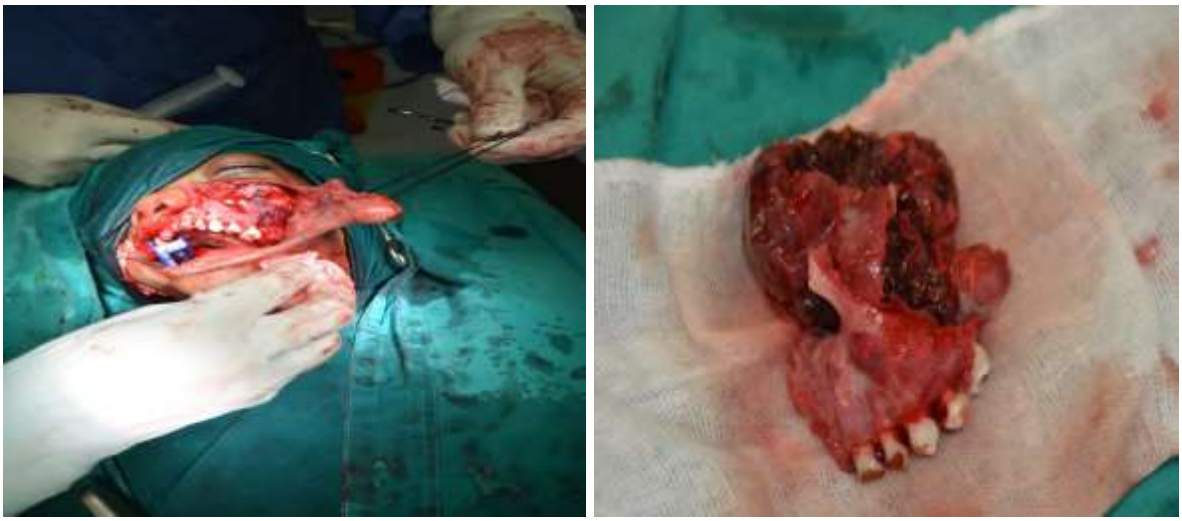

Fig (3):- Hemimaxillectomy through Weber-Ferguson incision.

Histopathology:-

Tumor tissue formed of immature bony trabeculae rimmed by osteoblasts in some areas embedded in cellular fibrous stroma fig (4), and multinucleated osteoclast-like giant cells was seen with occasional normal mitotic figures. Some psammomatoid bodies as basophilic bodies with esinophilic border were also detected fig (5). Large cavities filled with blood with no endothelial lining rimmed by osteoclasts fig (6).With correlation with young age and aggressive course the diagnosis was PJOF associated with ABC.

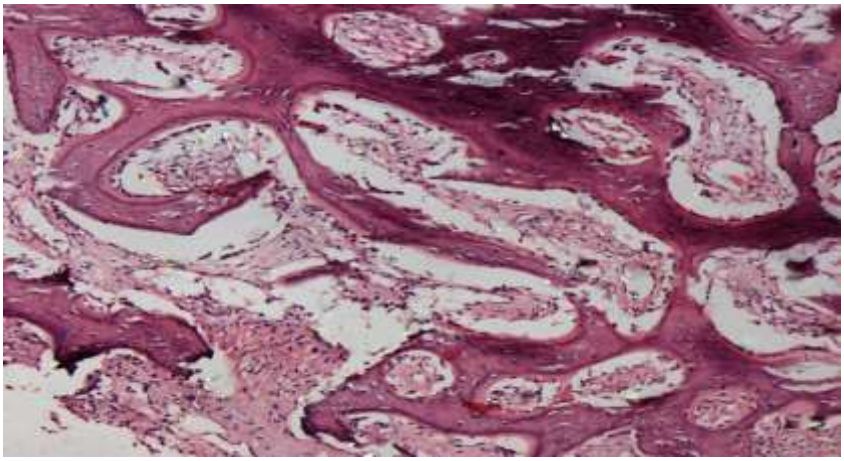

Fig (4):- bony trabeculae with variavle osteoblastic rimming in fibrous stroma.

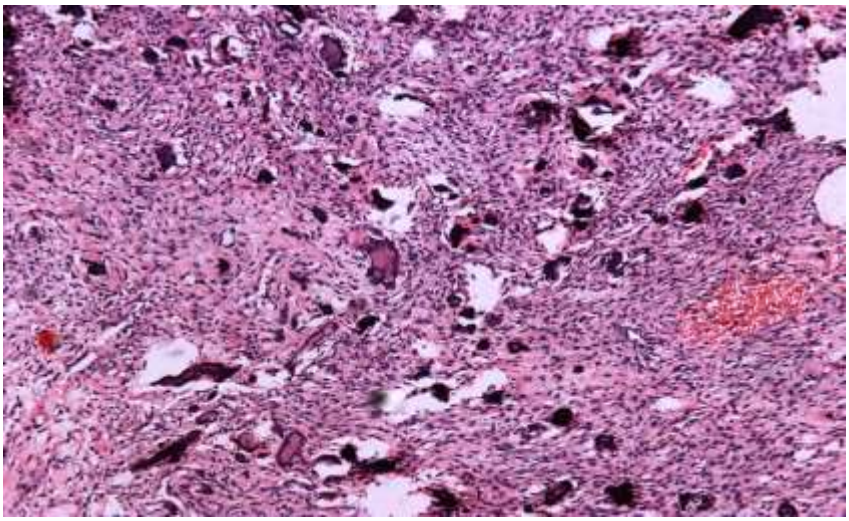

Fig (5):- multiple psammomatoid bodies with basophilic center and esinophilic periphery

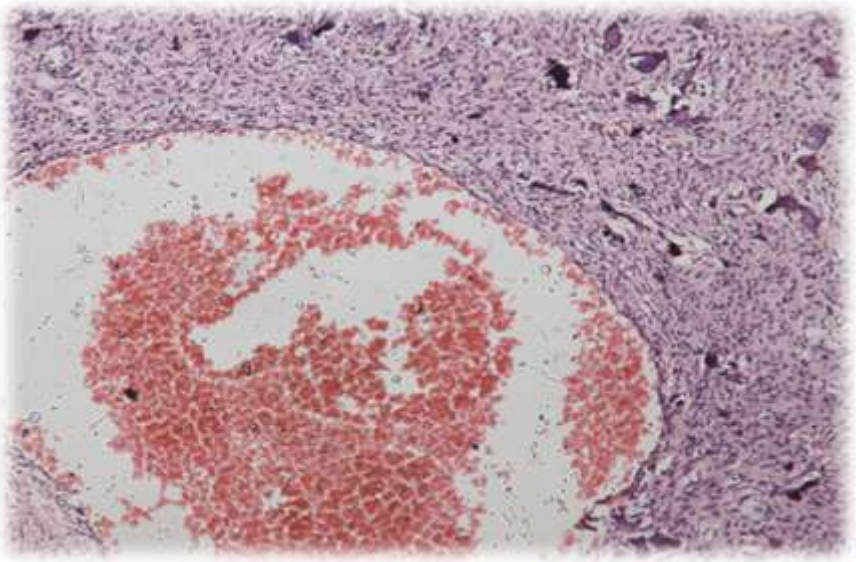

Fig (6):- large cavities filled with blood lacking endothelial lining representing ABC.

Discussion:-

PJOF has been considered as a distinct disease entity from conventional ossifying fibroma and the other fibro-osseous lesions because of its tendency to occur at a young age and its locally aggressive behavior. Moreover, PJOF may clinically manifest with rapid painless expansion of the affected bone as an aggressive lesion mimicking malignancy such as osteosarcoma or chondrsarcoma. So, it is critical to accurately recognize PJOF for making the proper diagnosis and management of this lesion without over emphasizing the situation (14).

The differential diagnosis in conjunction with radiographic findings in this patient initially included aneurysmal bone cyst, central giant cell granuloma, osteogenic sarcoma, and progressive monostotic fibrous dysplasia and nonodontogenic primary tumors of bone, such as osteoblastoma. In addition, locally aggressive odontogenic lesions, such as ameloblastic fibro-odontoma, as well as cystic lesions as calcifying odontogenic cyst and keratocystic odontogenic tumor. Morover vascular tumors can also have rapid growth and may be considered in the differential diagnosis. For example, central hemangioma grows rapidly and commonly presents as a radiolucent mass in children and young adults. Arteriovenous malformations also exhibit rapid growth, but usually display bruits on examination (14).

It is important to distinguish PJOF from central cementifying fibroma which is a benign jaw lesion of odontogenic origin arising in the molar and premolar regions of the maxilla with average age range in the third or fourth decade of life with a distinctly female predilection (3.4, 7). Radiographically, central cementifying fibroma is a well-marginated, unilocular, radiolucent or variably opacified lesion. Microscopically, central cementifying fibromas consist of fibrous stroma with dense cellularity and small, spherical calcifications "cementicles". Unlike PJOF, central cementifying fibromas has very low recurrence rate (7).

Histologically, psammoma-like bodies in PJOF were found to possess a dark rim of crystals from which small spicules and needle-like crystalloids project toward the periphery (21). The stroma varies from being loose and fibroblastic to intensely cellular with minimum intervening collagen. PJOF contains deeply basophilic concentrically lamellated particles as well as irregular thread-like or thorn-like calcified strands in a hyalinized stroma. In contrast to fibrous dysplasia, osteoclasts and osteoblasts typically line the trabeculae, which are composed of entrapped lamellar bone. Other features such as trabeculae of woven bone as well as lamellar bone, pseudocystic stromal degeneration, and hemorrhages result in areas similar to an aneurysmal bone cyst (2).

Treatment of PJOF has been controversial. Complete removal by enucleation alone or with curettage as well as en block resections are common therapeutic options. Most cases require tissue removal as much as possible while protecting adjacent vital structures to prevent recurrences. Our case required hemimaxillectomy to ensure total excision of the mass. The reported recurrence rate ranges between 30 and 58 % (4, 23). Radiotherapy has been proven ineffective and contraindicated due to an increased incidence of malignant transformation ranging from 0.4 to 40%. Despite the aggressive behavior, no metastasis has been reported (11). Therefore, conservative treatment of the jaw lesions by enucleation and curettage has been reported to be successful in non-aggressive cases.

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