Juvenile Psammomatoid Ossifying Fibroma of Maxillary Sinus: Case Report with Review of Literature

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Abstract: Juvenile psammomatoid ossifying fibroma is a rare benign fibro-osseous tumor of the gnathic and extra-gnathic craniofacial bones, particularly the periorbital, frontal and ethmoid bones. It is slowly progressive with aggressive local growth, invasion and destruction of the surrounding tissue, bone erosion and recurrence after surgical excision. It is distinguished from the other fibro-osseous lesions by its age of onset, clinical presentation and aggressive behavior.

Keywords Cemento-ossifying fibroma · Psammomatoid juvenile ossifying fibroma · Trabecular juvenile ossifying fibroma · Psammomatoid ossifying fibroma

Introduction

Psammomatoid ossifying fibroma (POF), also known as juvenile ossifying fibroma (JOF), and fibrous dysplasia of the jaw are maxillofacial lesions that should be distinguished from each other because they show distinguished pattern of disease progression [1]. However, both lesions

often show similar histological and radiological features, making distinguishing between the two, a diagnostic dilemma [1]. These lesions may be associated with significant cosmetic and functional disturbances. Pathognomonic histologic feature of POF is the presence of calcified psammomatoid bodies with concentric laminated appearance [2]. Because of its risk of recurrence and aggressive nature, ossifying fibroma needs to be detected early and completely enucleated followed by long term followup [3].

Case Report

A 14 year male patient reported to the Department of Oral and Maxillofacial Surgery of M. S. Ramaiah Dental College with a history of swelling on the left side of the face since 1 year, which was associated with epiphora, episodes of epistaxis and left nasal obstruction. The swelling gradually increased to the present size since 1 year with proptosis of the left eye, however the vision was normal. Before reporting to us the patient visited few other hospitals where no treatment was given and was referred elsewhere. The patient was admitted and pediatric, ophthalmology, otolaryngology, neurosurgery opinion was sought. There was no vision problem or similar swelling elsewhere in the body, there was no weight loss/no loss of appetite, no headache or any other central nervous system findings. The patient was poorly built and malnourished. On local examination, there was gross asymmetry of the face on the left side due to swelling which was irregular and diffuse, extending from the floor of the orbit downwards up to the corner of the mouth to the tragus. Medial limit of the swelling was within the nasal cavity which led to deviation of the septum. Posteriorly, the swelling was about 5 cm anterior to the tragus. On palpation there was a hard bony

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swelling confirming the inspection findings. On intraoral examination there was obliteration of the left buccal sulcus from 23 to 26, which on palpation yielded a cystic consistency of the swelling.

The computed tomography (CT) of the patient reported a expansile lytic lesion involving the left maxillary sinus, the floor of the orbit and the medial wall causing pressure effect and proptosis of the left eyeball. The fibro-osseous component was extending into the sphenoid sinus, the oropharynx and the nasopharynx. The expansile lesion was extending into the nasal cavity with narrowing of the right maxillary ostium. Retained secretion was seen in the ethmoid sinus.

The MRI of the patient reported a large expansile heterogeneous signal intensity lesion with hyperintense in T2 and T1RM sequence and hypointense in T1 sequence extending medially in the nasal cavity causing significant deviation of the nasal septum and extension of the tumor into the nasopharynx. Superiorly the lesion was extending into the floor of the orbit. There was mild narrowing of the orbital apex.

Incisional biopsy was taken from the intraoral site and blood tinged cystic fluid was aspirated. The biopsy was reported as JOF. The patient was planned for resection of the tumor and reconstruction with titanium mesh. Through the Weber Fergusson incision with infraorbital lynch extension, the lesion was exposed. A Lefort I osteotomy was performed to access the maxillary, nasal and nasopharyngeal extension of the tumor. Osteotomies through the body of zygoma, at the frontal process of the maxilla, pyriform aperture to the canine tooth region were performed. Tumor was “shelled out” completely from the skull base, from the oropharynx and nasopharynx along with the lateral wall of the nose. The left nasal bone was osteotomised and repositioned. The temporoparietal flap was raised and the temporalis muscle was utilized to support the globe. The down fractured maxilla was plated back into its original position. The defect was reconstructed using titanium mesh which was contoured to form the floor of the orbit, the malar prominence and the frontal process of the maxilla. Partial alveolectomy was performed in the region of canine and premolar area. The wound was closed in layers. The tissue was sent for biopsy. The postoperative period was uneventful. The patient is on regular follow up and no sign of recurrence was noted so far (Figs. 1–4).

Discussion

The spectrum of fibro-osseous lesions includes a variety of developmental, reactive or dysplastic lesions and neoplastic entities [4]. According to WHO classification in 2005, benign fibro-osseous lesions in the oral and maxillofacial

Fig. 1 Preoperative photograph of the patient

Fig. 2 Post-operative photograph of the patient

regions were divided into two categories, osteogenic neoplasms and non-neoplastic bone lesions; cemento-ossifying fibroma was included into the former category [5]. JOF constitute uncommon benign neoplastic entities. In these lesions, the normal bone architecture is replaced by fibroblast and collagen fibres containing various amounts of mineralized material [6]. Cemento-ossifying fibroma in the extragnathic facial bones are called POF. Johnson et al., in 1952 used the term juvenile active ossifying fibroma for POF in either the gnathic or extragnathic bones [7].

Psammomatoid juvenile ossifying fibroma (PsJOF) was reported by Benjamin in 1938 who gave it the designation, osteoid fibroma with atypical ossification of the frontal sinus. It was later termed POF of the nose and paranasal sinuses by Gogl in 1949 [4]. The most characteristic feature of JOF as the name suggests, is its higher incidence in children and young adults [8, 9], however it can occur in

Fig. 3 CT (axial section) showing lytic lesion involving the maxilla, nasopharynx

Fig. 4 CT (coronal section) showing lytic lesion involving the maxillary sinus, the body of zygoma, the alveolar process of maxilla

older age groups. Johnson et al. have reported JOFs occurring at any age from 3 months to 72 years (Figs. 5–8).

Slootweg et al. classified JOF into two distinct groups, the PsJOF (psammoma like ossicles) type and the trabecular JOF (TrJOF) type [4]. JPOF has got unique features in terms of age of occurrence, location and clinical features. Approximately 75 % of PsJOFs develop in the orbit, paranasal sinuses and calvaria, whereas TrJOF is predominantly a gnathic lesion affecting the jaws with predilection for the maxilla. Our patient had involvement of the left maxilla and maxillary sinus, sphenoid sinus, ethmoid sinus, oropharynx, nasopharynx and the nasal cavity (Figs. 9, 10).

Fig. 5 Photomicrograph showing multiple calcified "Psammomatoid bodies," H&E $\times 100$

Fig. 6 Photomicrograph showing eosinophilic ossicles with concentric laminations, H&E $\times 400$

Fig. 7 Intraoperative photograph showing the tumor mass through the Weber Fergusson incision

Fig. 8 Intraoperative photograph showing Lefort I access osteotomy

Fig. 10 Intraoperative photograph showing the reconstruction of the defect with titanium mesh

Fig. 9 Intraoperative photograph showing the defect after en masse excision of the tumor

JOF usually manifests as an asymptomatic bony hard swelling whose extent and duration may depend on the site of occurrence and aggressiveness of the lesion. It can expand the involved bone and cause gross asymmetry of the face [8]. In the maxilla, the tumor may lead to nasal obstruction, epistaxis, and eye displacement. Both types of JOFs have male predilection. In our case the patient was a young male with all the above symptoms.

Complications arising secondary to the tumor are results of impingement of the tumor on the neighboring structures. Margo et al. reported that the orbital involvement can be either directly or secondary to involvement of the paranasal sinuses. In our case the lesion was extending from the

maxillary sinus to the floor of the orbit causing proptosis and epiphora; however there was no change in vision [4].

Intracranial extensions have been seen in lesions in the vicinity of the cribriform plate. In our case the lesion on the CT image appeared to be close to the cribriform plate. Primary extra cranial psammomatoid meningioma (PEPM) constitutes 2 % of all PEPs and many of them involve paranasal sinuses and the cribriform plate of ethmoid. PEPs represent diagnostic challenge in this location as symptoms and radiological aspects of these tumors may be similar to those of JOFs [10]. In our case neurosurgical opinion was sought, MRI was done which ruled out involvement of the cranium, hence PEP was excluded.

In our case, radiographic differential diagnosis included conventional ossifying fibroma, fibrous dysplasia and mixed odontogenic tumors. PsJOF often reveals aggressive lesions with cortical destruction, which can be used to discriminate it from ossifying fibroma. Fibrous dysplasia manifests as ill-defined contiguous lesions because of its blending with normal bone, with expansion of the involved bone throughout its overall length over several years, which demonstrates a slow growth pattern [10, 11].

Boysen et al. stated that the differential diagnosis of fibrous dysplasia versus ossifying fibroma rests on a radiographic criterion after the histopathologist has verified the fibro-osseous nature of the lesion [7]. In our case, the differential diagnosis based on clinical manifestations and conventional radiographic studies was controversial. Histologic interpretation was critical and led to correct treatment.

CT imaging plays a major role in detecting the extent of such lesions, their diagnosis and planning the management

[12]. The CT of our patient reported expansile lytic lesion involving the left maxillary sinus, the floor of the orbit, the medial wall of the orbit causing pressure effect and proptosis of the left eyeball. The fibro-osseous component was seen extending into the sphenoid sinus, the oropharynx and the nasopharynx. The expansile lesion was extending into the nasal cavity displacing the nasal septum, obliterating the nasal cavity and compressing the contralateral maxillary sinus ostium. Retained secretion in the ethmoid sinuses was also noted.

The mixed odontogenic tumors, odontoma and ameloblastic fibroodontoma also occur in younger patients and present as mixed density mass lesions in the tooth bearing portions of the jaws. These tumors are well demarcated, often intimately associated with an unerupted tooth and contain coarse opacities that are similar in radiodensity to tooth structure [10].

Johnson et al. revealed cystic changes in 53 % of cases and the majority of the cysts were small. The cysts tend to occur more commonly in the younger patients in the first and second decade of life. Large aggressive maxillary lesions were commonly associated with aneurysmal bone cyst formation [13]. In our case cystic changes were also seen, about 3 ml of blood tinged cystic fluid was aspirated which on cytologic evaluation reported cystic proteinaceous material.

The origin of ossifying fibroma was thought to be from the periodontal membrane. Johnson et al. hypothesized that JPOF originates from overproduction of the myxofibrous cellular stroma normally involved in the growth of the septa in the paranasal sinuses as they enlarge and pneumatized. These stromal cells secrete hyaline material that ossifies and connective tissue mucin that initiates the cystic areas. A recent study by Sawyer et al. demonstrated the presence of non-random chromosome break points at Xq26 and 2q33 resulting in translocation [10].

Histologically, the juvenile variants share a similar stroma but PsJOF is characterized by innumerable small spherical/lamellated ossicles resembling psammoma bodies of meningiomas set in cellular fibrous tissue while TrJOF contains trabeculae of woven bone with coarse lacunae, swollen osteocytes and a lining of plump osteoblasts [14]. The separation of the two types is only on the basis of histological examination. PsJOF and TrJOF seem to behave similarly however and their management is the same.

Sopta et al. commented that in lesions with longer case history, the number of “Psammomatoid bodies” were greater. In our case there were a good number of psammoma bodies which coincided with the duration of the lesion. The histopathologic finding, radiographic and clinical finding confirmed the diagnosis of PsJOF in our case [15].

Management

JPOF is a rare benign fibro-osseous tumor of craniofacial bones, particularly the periorbital, frontal and ethmoid bones. It is slowly progressive with aggressive local growth, invasion and destruction of the surrounding tissue, bony erosion and has shown recurrence after surgical excision.

The aggressive nature of this lesion along with reports of high recurrence rate of 30–58 % suggests that PsJOF should be treated very much like ameloblastoma, i.e., surgical resection is the line of treatment rather than conservative curettage [8].

The preferred treatment for PsJOF is complete surgical excision and partial or incomplete resection leads to recurrences. The tumor mass should be removed en masse to the level of normal bone with preservation of adjacent vital structures. Reconstruction procedure can be carried out either as a primary or secondary procedure [10].

The entire tumor should be shelled out completely during the surgery. However maxillary lesions are more difficult to remove completely because of the quality of bone and larger size of the lesion at the time of presentation [16]. However in our case the lesion was well encapsulated and was completely shelled out from below the cribriform plate, the oropharynx, and nasopharynx without any remnant being left behind.

Prognosis

Prognosis is considered to be good, despite the lesions tending to locally invade and recur. There are no reported cases of metastasis. Recurrence after surgical management is common and is reported to range from 30 to 56 %. Of the 112 cases reviewed by Johnson et al., 30 % recurred one or more times. Aggressive growth was observed only in a minority of cases, typically occurring in children and in maxillary lesions. Aggressive behavior of the lesion could not be predicted histologically. Recurrence may be attributed to the difficulty in proper resection caused by the location of the lesion and the infiltrative nature of the tumor borders. In our case there has been no recurrence till date.

Conclusion

POFs represent unique subset of fibro-osseous lesions of the sinonasal tract. They have distinctive histomorphologic features and a tendency towards locally aggressive behavior including invasion and destruction of the adjacent anatomical structures. Complete surgical removal at the earliest

possible stage is the treatment of choice. Small lesions can be treated conservatively by curettage/enucleation. An open surgical approach is ideal for resecting large and irregular shaped tumors that infiltrate sinuses and frontonasal bones.

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