Juvenile Trabecular And Psammomatoid Variant Of Ossifying Fibroma Of The Maxilla With Secondary Aneurysmal Bone Cyst.

L Emerson., M Alexander, A Job

Citation

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Abstract

Juvenile ossifying fibroma (JOF) is a locally aggressive benign lesion most commonly involving the facial bones. It has been classified as psammomatoid (Ps JOF) and trabecular (Tr JOF) based on histological criteria. Both the variants are known to have overlapping histopathological features with other variants of ossifying fibromas. Complete surgical excision is the treatment of choice. Here, a case of JOF with histopathological features of both psammomatoid and trabecular pattern with secondary aneurysmal bone cyst is presented along with clinical and radiological findings.

INTRODUCTION

Juvenile ossifying fibroma is an aggressive lesion with higher morbidities associated with lesions involving head and neck region. Depending on the site, it can cause facial asymmetry, pain, paresthesia, malocclusion and proptosis. Histopathologically, it has been classified into two variants, Trabecular and Psammomatoid by El-Mofty S [1] Overlap of the variants has been found to be associated with other variants of ossifying fibroma [2] Both variants have been associated with different age groups.

Whether both variants are part of single entity is still a matter of debate [3] Bohn OL et al report a variant of JOF, mimicking Psammomatoid Meningioma [4] Treatment is complete surgical excision. Recurrences are reported to be treated with chemotherapeutic agents [5] in addition to surgery.

CASE REPORT

A 16-year-old male presented with complaints of obstruction of the left side of nasal cavity for the past six months. Nasal obstruction was progressive, leading to mouth breathing. It was associated with swelling of the left side of the face and proptosis of the left eye. There were associated symptoms of anosmia and multiple episodes of epistaxis. There was no loosening of teeth. There was no prior history of trauma or any other etiology, which could contribute to the pathology of the lesion.

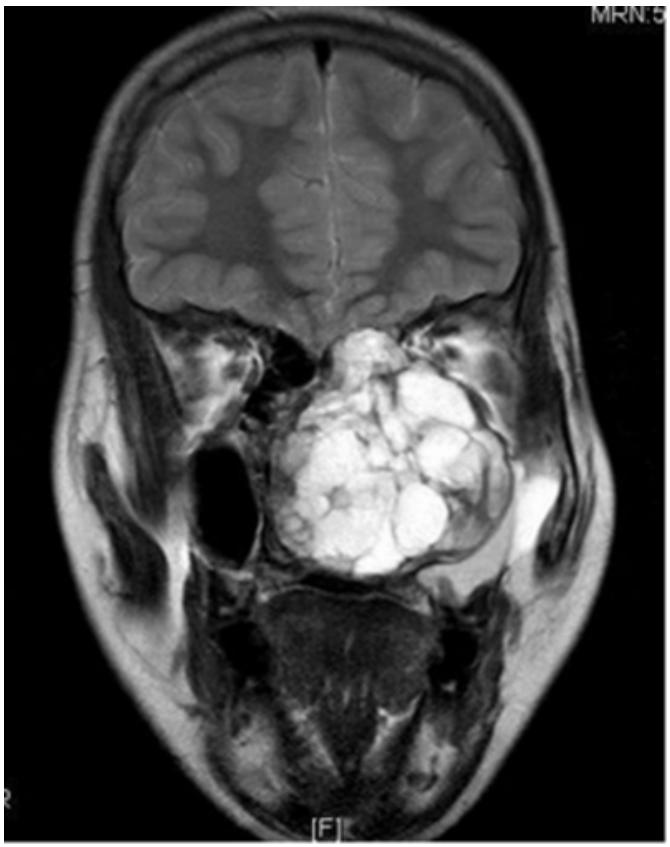
On examination, the external osseo-cartilaginous framework was altered with splaying of the nasal cartilages and loss of nasolabial fold on the left side. Anterior rhinoscopy revealed a pink polypoidal mass, which was firm, filling the left nasal cavity. There was contralateral deviation of the nasal septum due to the pressure of the mass. The mass was found to extend into the naso-pharynx.

There was no swelling in the oral cavity and the teeth were intact. Mild proptosis of the left eye was noted with restriction of movements laterally and superiorly. An ophthalmology consult revealed normal vision with restriction of ocular movements on the lateral gaze.

A preliminary biopsy of the mass done elsewhere showed features of benign lesion.

Figure 1

Fig-1. T2 weighted mri image showing multiple locules with compression of orbital structures



A CT Scan revealed a large, 6.5(AP) x 5(CC) x 6(Tr) cm, well-defined, multi-loculated soft tissue density lesion with expansion of the surrounding bones with remodeling. A MRI with contrast(Fig-I) showed predominantly long-TR hyperintense mass with hypointense septations in the left nasal cavity and ethmoid sinuses. Multiple fluid-fluid levels were seen within many of the locules. There was no intracranial extension of the mass.

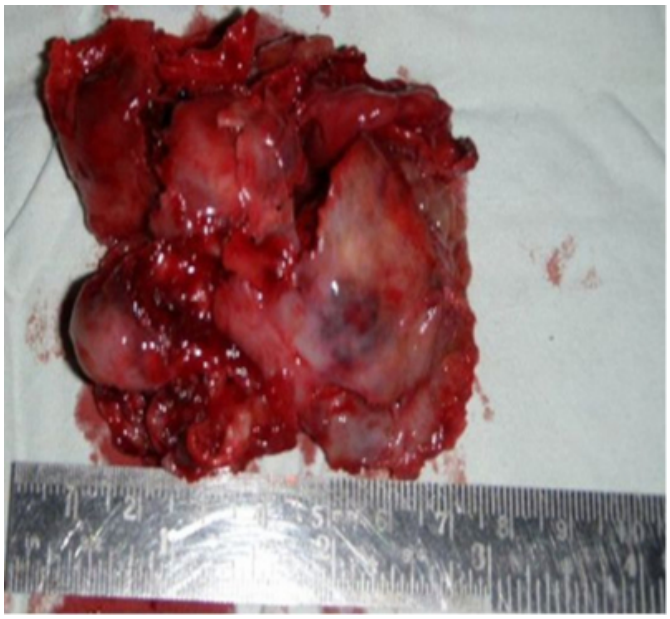
A differential diagnosis of ossifying fibrous lesion or aneurysmal bone cyst was made. The lesion was removed by a lateral rhinotomy approach. The mass was found to be loculated with a thin shell of bone with erosion of the medial wall of the orbit compressing the orbital apex. The mass was abutting the cribriform plate with considerable thinning of the plate. The mass was excised in total. There was no CSF leak.

The specimen (Fig -2) received was in multiple pieces of soft to bony dark brown tissue. The largest mass (150g) had measurement of 6.5 x 2 x 2cm. It was partially covered by a thin shell of bone. The external surface was hemorrhagic.

Sectioning of the largest piece revealed cystic spaces filled with brown granular material. Histologic sections were prepared from 10% formalin-fixed, paraffin-embedded tissue and stained with hematoxylin and eosin.

Figure 2

FIGURE-2: Gross view of the specimen



Hematoxylin and eosin-stained sections (4µ) from the mass showed a lesion composed of immature woven bone and mature lamellar bone trabeculae with and without osteoblastic rimming (Fig -3) with intervening fibrocellular stroma. The stroma was cellular at places and forming storiform sheets of densely packed spindle shaped cells with occasional mitotic figures and intervening “cementicles” psammoma-like material (Fig 4). Fibrocollagenous septae with large blood filled spaces, occasional multinucleate giant cells, areas of hemorrhage with changes consistent with secondary aneurysmal bone cyst and granulation tissue were also present. Foci of respiratory epithelium and mucous glands were present. These features (Fig-5) are consistent with the trabecular and psammomatoid variants of ossifying fibroma with secondary aneurysmal bone cyst.

Figure 3

FIGURE-3 showing trabeculae of lamellar bone with and without an osteoblastic rimming and intervening large blood filled spaces(ABC like changes) in fibrocellular stroma (H&E section, 200X)

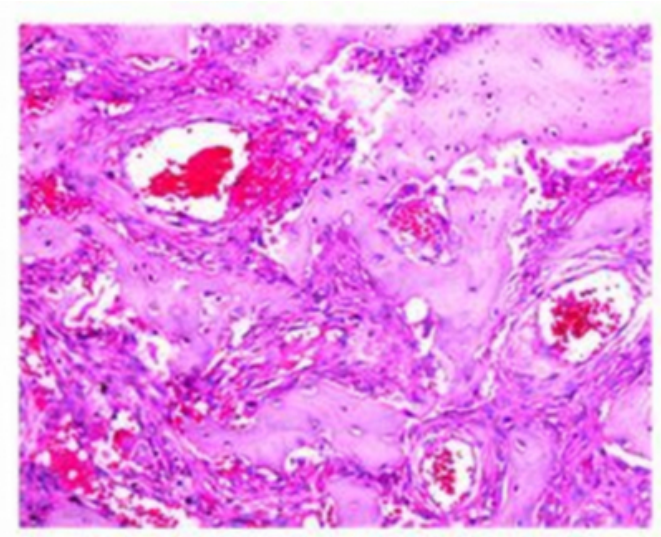


Figure 4

Fig.4 showing psammoma like “cementicles” intervening sheets of spindle cell stroma (H&E section,400X)

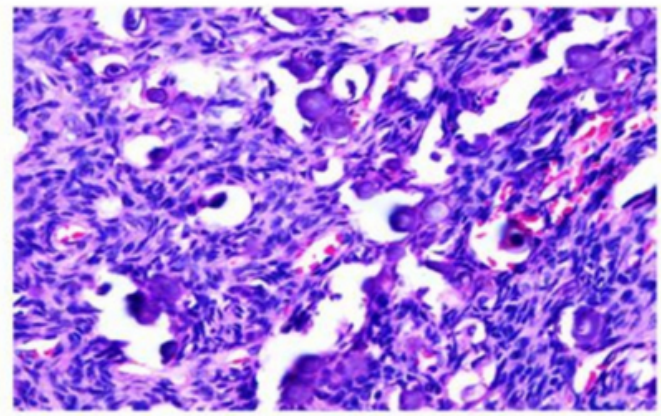
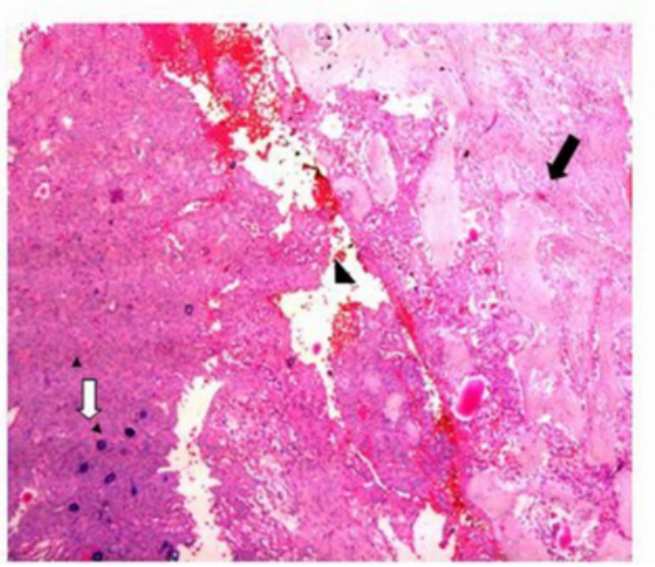


Figure 5

FIGURE-5: Showing all the three features: Woven Bone Trabeculae (BLACK ARROW) with intervening fibro cellular stroma, large blood filled spaces (Arrowhead) and sheets of spindle cells with cementides (White arrow) in between. H&E SECTION 100X



DISCUSSION

Fibro-osseous neoplasia is a generic term encompassing several conditions. Menzel first reported ossifying fibromas in 1872. Montgomery [6] gave its description as a separate entity. Due to its aggressive nature in children and characteristic histopathological features, it was separated from the group of ossifying fibroma. Batsakis [7] considers them examples of one disease entity. Makek [8] proposed a ‘working’ classification of fibro-osseous lesions. The nomenclature for the group involving nasal cavity and maxillary sinuses includes ossifying fibroma, cementifying or cemento-ossifying fibroma, psammomatoid ossifying fibroma and juvenile active ossifying fibroma. Juvenile ossifying fibroma (JOF) is considered a distinct entity because of its tendency to occur in young age groups and due to its locally aggressive nature. JOF was defined by Reed and Hagy as ‘a localised actively growing destructive lesion occurring predominantly in children and characterised histologically by trabeculae of woven bone showing focal lamellar bone replacement in a cellular stroma’ and may mimic a malignant neoplasm [9] . It was classified into two separate subtypes as psammomatoid and trabecular ossifying fibroma. According to El-Mofty, the trabecular variant is seen more commonly in ages 8-12 years, and the psammomatoid variant in 16-33 years. The most common

sites in the head and neck region [10] are the maxilla, ethmoid and the frontal bones, although it can occur anywhere in the skeleton. Microscopically, Tr JOF consists of a mixture of cellular osteoids without osteoblastic rimming and trabeculae of immature bone with osteoblastic rimming. Ps JOF shows spherical or ovoid ossicles that resemble psammoma bodies with or without osteoblastic rimming. Secondary changes such as aneurysmal bone cysts and hemorrhage are commonly associated with JOF. Aggressiveness was attributed to the extremely cellular fibrous stroma in JOF [11] The "aggressive" biologic behavior of the "juvenile" variants appears to be a consequence of increased likelihood to encounter recurrences in sinonasal-based tumors initially treated conservatively. Ossifying fibromas, usually multiple, are a common component of rare hyperparathyroidism-jaw tumor syndrome, in which there is a mutation of the endocrine tumour gene, HRPT2, on chromosome 1q encoding parafibromin. "Recurrent" tumors may represent development of a new ossifying fibroma rather than persistent/recurrent disease. The juvenile trabecular variant is a mimic of osteosarcoma because of bands of cellular osteoid and trabeculae of immature bone that may form a lattice, as well as the presence of mitotic figures. [12]

There are no reports of metastatic disease, however in view of its locally aggressive nature and tendency for recurrence, surgical resection has been advocated for.

This case report is presented to highlight the pathological controversy, which still surrounds this entity.

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Author Information

L. Paul Emerson., Bsc. MBBS, DLO, MS (ENT)
Department Of ENT, Unit-I, Christian Medical College Vellore

Manika Alexander, MBBS, M.D (PATH)
Department Of Pathology, Christian Medical College Vellore

Amand Job, MBBS, DLO MS (ENT), MNAMS
Department Of ENT, Unit-I, Christian Medical College Vellore