Case Report

Psammomatoid Ossifying Fibroma of the Ethmoid Sinus with Secondary Intracranial Aneurymal Bone Cyst: A Case Report and Literature Review

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A B S T R A C T

Juvenile psammomatoid ossifying fibroma (JPOF) is a rare, slowly progressive tumor of the extragnathic craniofacial bones, with a tendency toward locally aggressive behavior and recurrence. The pathognomonic histopathologic feature is the presence of spherical ossicles, which are similar to psammoma bodies. Very few cases in association with secondary aneurysmal bone cyst (ABC) formation have been reported in literature. Treatment consists of complete surgical removal. However, incomplete excision has been associated with a high local recurrence rate. The prognosis is good because malignant change and metastasis have not been reported. The authors are reporting a case of JPOF of the ethmoid bones with secondary ABC in a 7-year-old female patient.

Key words: Aneurysmal bone cyst, endoscopic management, paranasal sinuses, psammomatoid ossifying fibroma

ملخص البحث :

الورم الليفي الرملي العظمي من الحالات النادرة وهو ورم بطئ النمو داخل الجمجمة وذو طبيعة متكررة. يعرض الباحثون حالة لطفلة تبلغ من العمر 7 سنوات تم تشخيصها بهذا الورم في الجيب ألغربالي داخل الجمجمة. وتم علاجها جراحيا باستئصال الورم كاملاً باستخدام المنظار.

INTRODUCTION

Ossifying fibroma (OF), specifically the psammomatoid type, is an example of a benign, expansible, locally aggressive tumor of the extragnathic craniofacial bone. It can be associated with secondary changes, including aneurysmal bone cyst (ABC) like changes, myxoid changes, and hemorrhage. It consists of multiple blood-filled spaces of variable size that are separated by connective tissue.

These tumors rarely involve the sinonasal tract, and ethmoid sinus involvement is very rare. Typically, psammomatoid ossifying fibroma (POF) manifests macroscopically as a cystic tumor and microscopically, with the presence of spherical ossicles, looks like psammoma bodies.[1,2]

Here, we present a case of juvenile psammomatoid ossifying fibroma (JPOF) with secondary ABC of the ethmoid bone with intracranial extension in a 7-year-old female patient. A review of the literature revealed that this is the fourth case in which the paranasal sinuses are involved and the second case which was managed solely through a transnasal endoscopic approach.

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CASE REPORT

A 7-year-old girl was referred to the ENT clinic by neurosurgery as she had been suffering from a painless, gradually progressive proptosis of the left eye for 6 months. Her vision was normal, and there was no history of nasal obstruction, trauma, or epistaxis. General and systemic examinations were normal. Transnasal endoscopic examination revealed the mass was hard and smooth covered with normal mucosa filling the left middle meatus. There was obvious axial proptosis, but ocular movements were normal, and pupillary reaction and corneal sensation were intact. Fundus examination and routine blood serum electrolytes, chest X-ray, and urine analysis were normal.

Computed tomography (CT) of the nose and paranasal sinuses showed a well-defined expansile lesion of the mid ethmoid and frontal sinus with extensive bone remodeling and thinning of the adjacent lamina papyracea and superior orbital roof, with internal heterogeneous soft tissue component. Intracranial extension had a distinct cleavage line formed by thin bone separating the dura from the cyst component [Figure 1]. The magnetic resonance imaging (MRI) showed concentric soft tissue and multiple fluid levels of high signal intensity on T1 and T2 sequences with mild enhancement post contrast administration [Figure 2]. The carotid angiogram revealed no significant uptake [Figure 3].

The patient underwent transnasal endoscopic approach with navigational guidance in the presence of a neurosurgeon. There was a hard and smooth mass with a bony shell covered by an intact mucosa in the left nasal cavity within the middle meatus, which pushed the middle turbinate medially and the lamina papyracea laterally.

The mass consisted of multiple bony shells and cysts filled with blood which was excised completely using the 30° and 45° endoscopes under navigational guidance, which assisted us in identifying and avoiding insult to vital structures such as the orbit and the skull base and also helped in confirming complete excision of the mass. The entire nasal and the intracranial component of the mass were removed endoscopically resulting in the intact thin bone separating the nasal cavity and the cranium [Figure 4]. The patient had an uneventful postsurgical course. The pack was removed on the first postoperative day, and she was discharged on the third postoperative day.

Histopathological examination of the excised mass revealed numerous small ossicles or psammomatoid bodies embedded in the cellular fibrous stroma [Figure 5]. Large cystic spaces filled with blood and separated by fibrous septa were seen focally. These cysts were lined by fibroblasts and histiocytes [Figure 5]. Clusters of osteoclast-like multinucleated giant cells with loose spindly to cellular stroma and reactive woven bone were also noted. No malignant osteoid or atypia were identified. These findings are consistent with POF with underlying ABC.

DISCUSSION

Juvenile ossifying fibroma is a benign fibro-osseous lesion with occurrence in the head and neck region uncommon. It was first described by Benjamin in 1938 as "osteoid fibroma with atypical ossification."[3] Then in 1949, the "psammomatoid fibroma of the nose and paranasal sinuses" was initiated by Gogl.[4] Clinically, because of

Figure 1: (a-c) Different cross bonding views (axial, sagittal, and coronal) bone window computed tomography image shows an elliptic-shape, well-defined, and mixed density mass with peripheral ground-glass density arising from the left ethmoid sinus and pushing the left orbit out and down word.

Figure 2: (a) Sagittal T1-weighted image shows isointense signal of peripheral ossifying fibroma (OF). (b and c) Peripheral remnant OF has hypointense signal, and aneurysmal bone cyst (ABC) shows multicystic, heterogeneous signal intensity with multiple fluid-fluid levels on the axial T2-weighted image. (d) Axial contrast-enhanced T1-weighted image with fat saturation shows moderate enhancement of OF and, septal and peripheral enhancement of ABC.

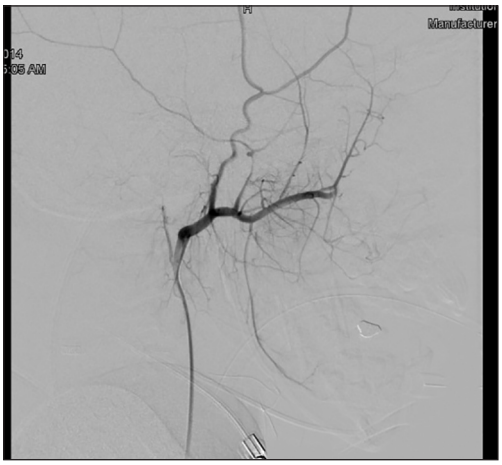

Figure 3: Left carotid arteriogram shows all the branches with no uptake by the mass.

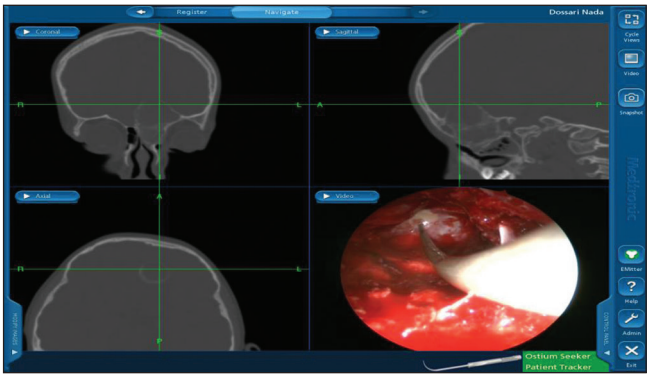

Figure 4: Intraoperative endoscopic view with navigation guided after the complete excision of the mass.

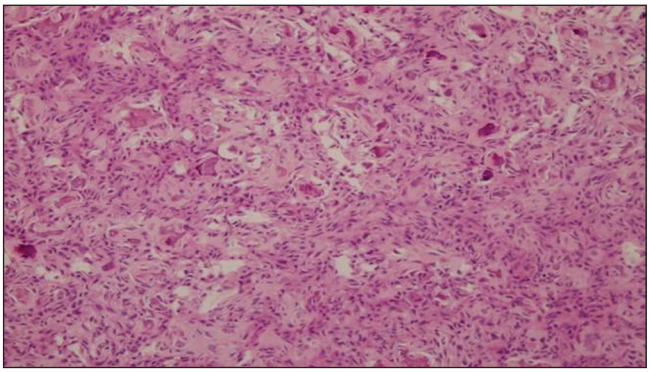

Figure 5: Microscopic picture shows the psammomatoid bodies embedded in a fibrous stroma (×100).

the aggressive nature of the tumor and its invasion into the surrounding structures, the term "Juvenile active OF" was given by Johnson *et al*. in 1952.[5] The presence of psammoma-like ossicles in juvenile OF was first described in 1994 by Slootweg *et al*.[3] In 2005, the World Health Organization further divided OF into two variants, namely juvenile trabecular OF and JPOF, based on the histopathological features.[4,5]

The clinical presentations of the OF depend on the site of involvement. Proptosis, diplopia, blindness, nasal obstruction, headache, and epistaxis are possible complaints. Although it can present at any age, from 3 months up to 72 years of age, it peaks in children and adolescents with slight male predominance (1.2:1).[4-7]

Psammomatoid ossifying fibroma rarely occurs in the sinonasal tract, including the periorbital, frontal, and ethmoid bones. Our present case is a 7-year-old girl with left eye proptosis and a lesion which originated from the left ethmoid sinus. To the best of our knowledge, four cases of sinonasal tract OF with secondary ABC have been reported to date in the literature.

Radiological evaluation, including a CT scan and MRI, is needed to evaluate the extent of the disease as well as the adjacent anatomy. On a CT scan, the mass appears expansile and circumscribed by a thick shell of high density bone with a multiloculated internal form and a content which can be radiolucent, mixed, or radiopaque, depending on the degree of calcification.[2,4,5] On the other hand, an MRI that shows a cystic mass with an outer hypo-intense shell suggests osteoid matrix calcification.[4,8] It is hypointense to muscle on T1- and T2-weighted sequences. The bony wall is iso-intense with the gray matter on T1-weighted images and hypointense on T2-weighted images. The enhancement of the bony wall with gadolinium suggests that there is no reactive hyperostosis.[8]

Clinically, the differential diagnosis is wide and includes benign and malignant nasal diseases. In our patient, the mass was a non-destructive, slowly progressive mass which was confirmed by benign pathology, such as fibrous dysplasia, cementoblastoma, and mucocele. A definitive diagnosis was made by histopathology.

Microscopically, POF can mimic psammomatous meningioma involving the craniofacial bones. Immunohistochemistry is not of great help, and the differentiation is solely based on the morphology.[9] The even distribution of the calcification, the true ossification, beside the inconspicuous meningothelial features such as the syncytial cells with nuclear pseudo-inclusions, should favor POF. Moreover, presentation at a young age, such as the presented case, does not support a diagnosis of meningioma.[10]

The ABC is an expansile osteolytic lesion, often multilocular, with blood-filled spaces separated by fibrous septa containing osteoclast-type giant cells and reactive

bone. The present case was characterized by large cystic areas of ABC showing giant cells and osteoid formation. Development of ABC in JPOF has been previously reported.[5,11,12] It develops initially as a focal myxoid change in the hemorrhagic stroma. Osteoclastic giant cells are seen with gradual expansion and formation of cysts that have thin fibrous walls. The cysts tend to occur commonly in young patients, predominantly in the first and second decades of life and are commonly associated with lesions involving the maxillary sinus.[5]

The advocated treatment for JPOF is complete surgical excision, as partial or incomplete resection might lead to recurrences.[7,13] The tumor mass must be removed down to the level of normal bone with preservation of adjacent vital structures as much as possible to avoid further potential complications due to the expansile nature of the mass and erosion of the surrounding vital structure.

The present case is the second case reported for the complete excision of POF with secondary ABC via the endoscopic transnasal approach, which is a safer procedure than the transcranial approach. The follow-up is recommended for such patients as lesions may reoccur. The more aggressive lesions have a recurrence rate of 30-56%.[1,5,6] Most of the recurrences seem to occur within the 1st year of surgical treatment.[14] It was recommended that this patient undergo a clinical endoscopic examination on a weekly basis for the 1st month followed by an examination every 2 months for the 1st year. Patients need to have the first radiological scan 2 months postsurgery to check for residual or recurrence unless the patient has a positive clinical finding, in which case the radiological exam should be done sooner.

CONCLUSION

Juvenile psammomatoid ossifying fibroma is a rare tumor of the extragnathic craniofacial bones. It is a slowly progressive lesion with a tendency to invade surrounding tissue and recur after surgical excision. Due to the possibility of the presence of hybrid lesions in this tumor, it is preferable to remove it en masse and take multiple sections for histopathological reporting. However, surgical approaches and techniques have not been well-defined, especially in young patients. With the invention of the navigation system, we can deal with such complicated cases more safely where complete surgical excision of the tumor is possible when surgery is based on preplanned criteria followed by clinical and radiological follow-up.

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Conflicts of interest

There are no conflicts of interest.

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