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Case Report

Transcranial resection of a juvenile psammomatoid ossifying fibroma of the orbit: A case report with 2-year follow-up

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ABSTRACT

Background: Juvenile psammomatoid ossifying fibromas (JPOFs) are benign, locally invasive lesion of the craniofacial skeleton that may undergo rapid growth resulting in damage to cranial and facial structures. They usually occur before the age of 15 years and should be carefully treated as their diagnosis may be confused with other lesions such as psammomatous meningioma.

Case Description: A 14-year-old male presented to the clinic with a history of progressive left proptosis. Imaging studies revealed a well-circumscribed lesion involving the left orbital roof and showing internal areas of calcification and sclerosis. He underwent a transcranial resection of the lesion and follow-up imaging revealed no evidence of recurrence.

Conclusion: JPOFs are locally invasive lesions that require careful diagnosis and meticulous excision to prevent recurrence.

Keywords: Fibro-osseous lesions, Juvenile ossifying fibroma, Orbit, Psammomatoid

INTRODUCTION

Juvenile psammomatoid ossifying fibromas (JPOFs) are among a group of ossifying fibrous lesions that also includes conventional ossifying fibroma (COF), JPOF, and juvenile trabecular ossifying fibroma (JTOF).[8] They are benign lesions with no reported malignant potential, however, they are sometimes subject to rapid growth that can make alarm bells ring. Their growth is due to replacement of bone by a fibrous cellular stroma with varying areas of bony trabeculae and cementum like material. The pattern of mineralization is what separates JPOF from its counterparts. The need for an accurate diagnosis and proper treatment is essential since these lesions may be locally destructive to the craniofacial skeleton.[1]

JPOF typically affects people younger than 15 years of age and usually arises in the bones of the paranasal sinuses, orbit, and frontoethmoidal complex. It is notorious for recurrence

following surgery and requires *en bloc* removal for complete extirpation. Radiologically, it is characterized by being a well-defined osteolytic lesion with a sclerotic rim.[3]

We present a case of a JPOF in a 14-year-old male who presented to our clinic with left sided painless proptosis. He underwent resection through a transcranial left frontal approach.

CASE PRESENTATION

A 14-year-old male patient presented to the outpatient department with a 3-month history of painless proptosis of the left eye [Figure 1]. The proptosis was progressive overtime but there were no visual symptoms on presentation. On examination, there were no cranial nerve deficits. General physical and rest of the neurological examination was unremarkable. CT scan of the head revealed a well-circumscribed lesion involving the left orbital roof and showing internal areas of calcification and sclerosis [Figure 2a-c]. MRI brain plain revealed the lesion to have a heterogeneous appearance with hyper to isointense areas. It was extra-axial and exerting mass effect over the base of the left frontal lobe [Figure 3]. Considering the growth of the lesion, it was decided to operate on it.

He underwent a left frontal craniotomy and subfrontal approach to the lesion with minimal retraction of the frontal lobe. The lesion was moderately vascular and firm in consistency. It appeared to be arising from the roof of the orbit and hence during excision, the periorbita was exposed which appeared to be disease free. Careful examination was performed to ensure maximum possible excision. An immediate postoperative CT with contrast showed complete removal of the lesion as well as the roof of the orbit [Figure 4].

The patient's recovery was uncomplicated and 1 year follow-up showed good scar healing. There was some residual ptosis with restriction of extraocular movements. Postoperative MRI brain with contrast [Figures 5a and b] demonstrates gross total resection. The patient remained on regular radiological follow-up. Histopathology was finalized as juvenile psammomatous ossifying fibroma [Figure 6]. Follow-up at 2 years with plain CT head showed no evidence of recurrence [Figure 7].

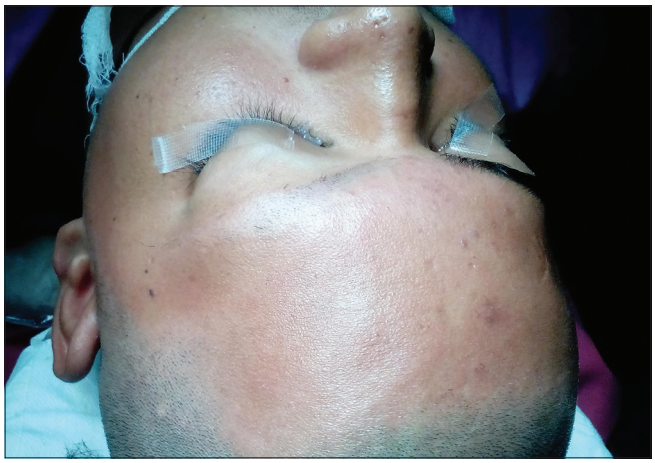

Figure 1: Preoperative photograph demonstrating left eye ptosis and positioning for a left frontal craniotomy.

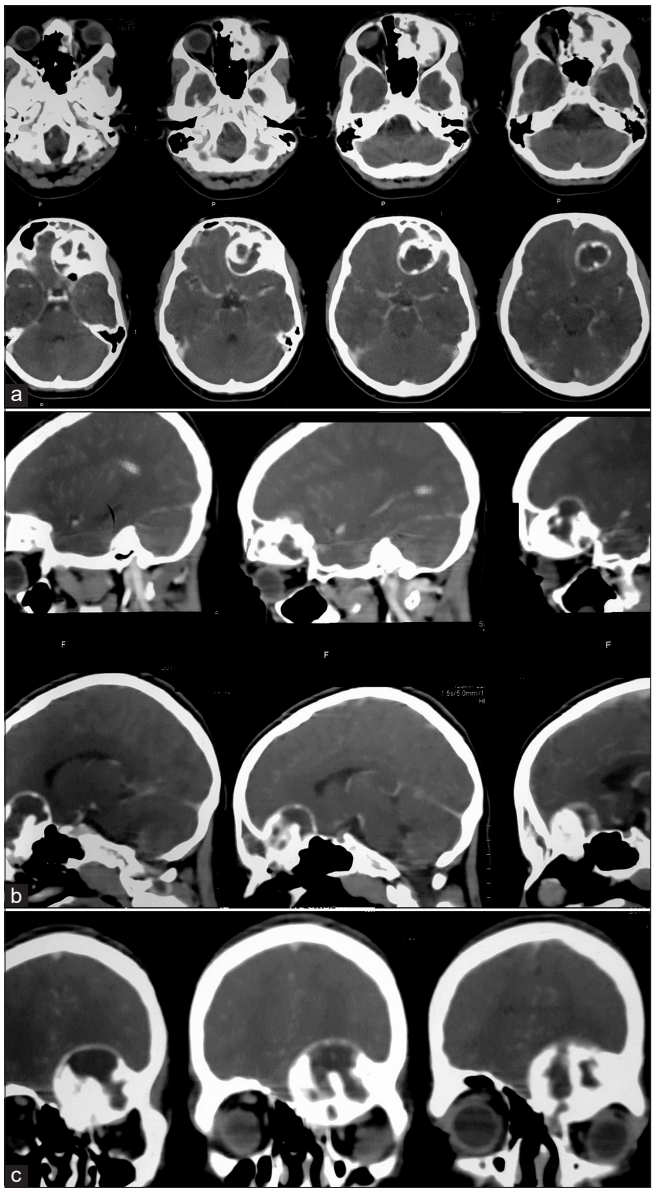

Figure 2: (a)Pre-operative CT brain with contrast axial sections demonstrating an anterior cranial fossa space occupying lesion with involvement of the orbital roof. There was a sclerotic contrast enhancing rim as well. (b) Preoperative CT brain with contrast sagittal sections demonstrating the lesion to be involving the orbital roof and causing mass effect and forward pushing the contents of the orbit. (c) Preoperative CT brain with contrast coronal sections through the orbit demonstrating the extent of the lesion to the midline and possible involvement of the cribriform plate.

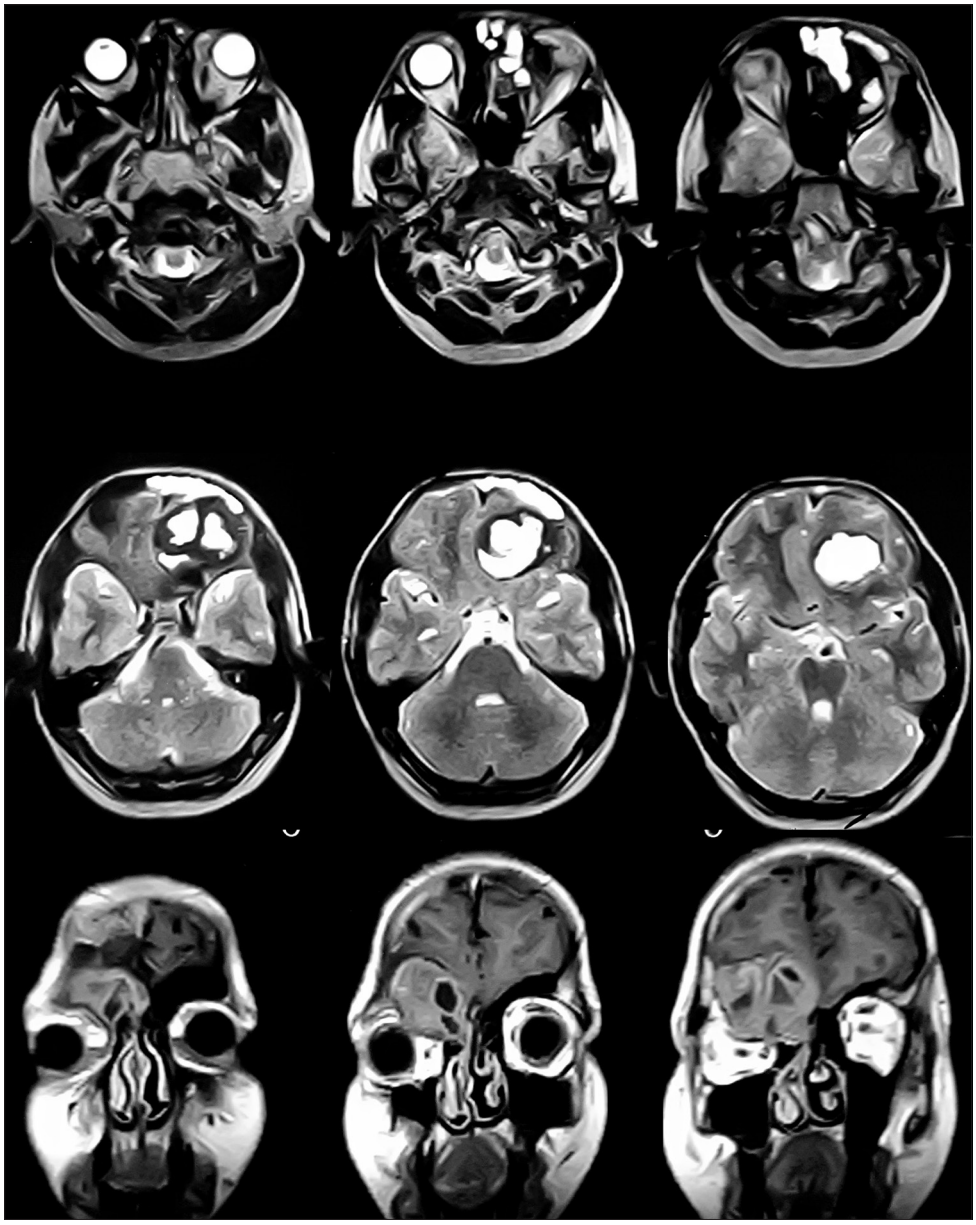

Figure 3: Pre-operative MRI Brain without contrast, T2 weighted axial images (top and middle row), demonstrating the extra axial nature of the lesion and mass effect on the frontal lobes with midline shift towards the right. T1 weighted post contrast coronal images (bottom row) demonstrates areas of heterogenous enhancement.

DISCUSSION

Some authors have recommended that a pretreatment diagnosis of JPOF be established before undertaking surgery. This is due to the difference in management and prognosis between the similar lesions in the group. JPOF mandates complete excision while other variants such as COF require simple curettage. Even histopathological similarities are present in the spectrum, and hence, a complete review of imaging and clinical history must be considered.[6,7]

JPOF can occur in either the craniofacial skeleton including the orbit and paranasal but never in tooth bearing areas such as the maxilla or mandible. They are generally unencapsulated and can have growth spurts with rapid local invasion and skeletal destruction.[2] Primary extracranial meningiomas are an important differential of JPOF as both can cause local bone changes and have a similar radiological appearance. This is more so true in the case of psammomatous meningiomas as they may have a similar immunotype as JPOF including EMA and vimentin positivity. Therefore, the diagnosis should be always include

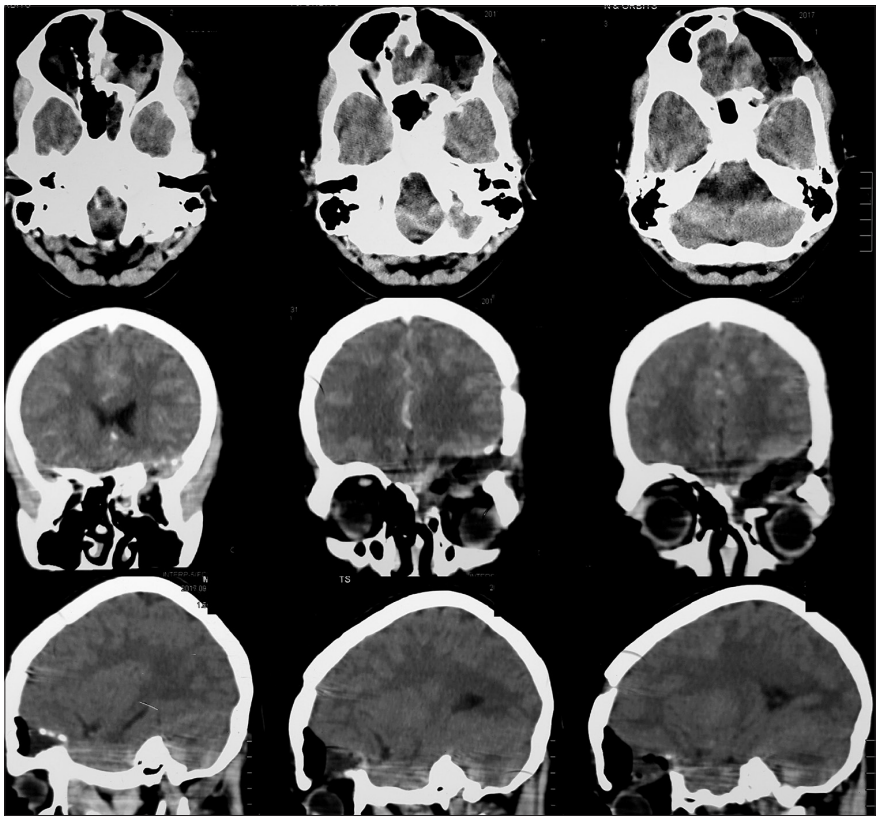

Figure 4: Post op CT head plain demonstrating interval resection of the lesion as well as the roof and medial wall of the orbit.

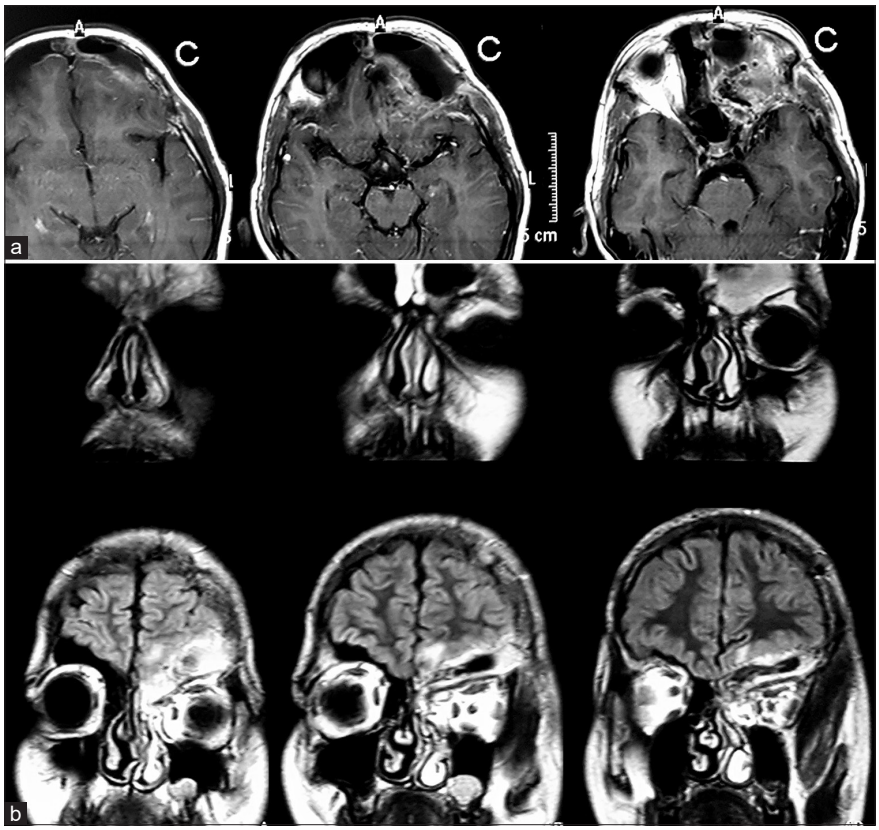

Figure 5: (a) Post-operative MRI Brain T1 post contrast axial section demonstrating interval resection of the lesion with resolution of mass effect and midline shift. (b) Post-operative MRI Brain post contrast FLAIR coronal images demonstrating the resection cavity extending to the midline.

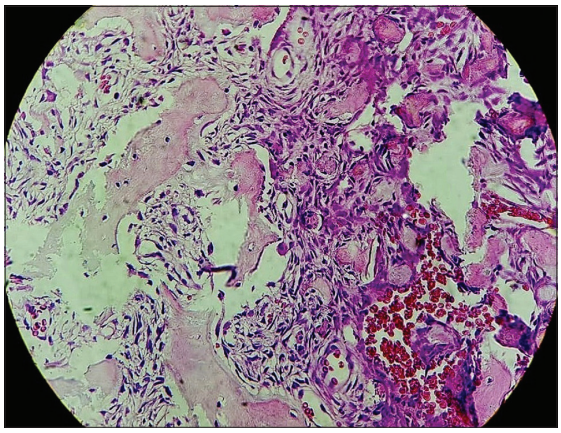

Figure 6: Microphotograph demonstrating numerous small, round ossicles (psammomatoid bodies) embedded in cellular fibrous stroma and within bony trabeculae.

a review of clinical and radiographic features as described by Noudel *et al*.[4]

Although prior confirmation through a biopsy before surgery is recommended by some authors, this may not always be possible as the lesion is rare and prone to being missed. We did not perform a biopsy in this case before attempting resection. Unfortunately, we do not have neuronavigation or frozen section available at our center and multiple surgeries would have placed significant financial pressure on the family. Hence, the plan of attempting maximum safe resection with available information was undertaken. Complete surgical excision should be the goal and is usually curative if successful. Neuronavigation may be useful in helping to achieve maximal resection. Recurrence following surgery has been reported to be upward of 50% and is due to incomplete excision. Complete excision may be challenging due to the complex anatomy of the regions involved and the ability of the lesion to involve and damage contiguous structures. In our case, the lesion involved the left supraorbital region and was amenable to an open approach that exposed the entire extent of the lesion. Wide open approaches may be preferred to ensure complete resection; however, several authors have reported that minimally invasive endoscopic approached may be advised. We would, however, express concern that since most of these tumors are firm to hard in consistency, an open approach with aggressive resection would be better. Patients may be followed up closely to detect any recurrence.[5]

Outcomes are good in cases of JPOF considering their benign nature but they can be fatal if untreated due to their unchecked growth. Cosmetic issues may also arise secondary to their growth and treatment. It is recommended that a multidisciplinary approach be used depending on the involvement of several areas of the face. In our case, the lesion was involving the roof of the orbit. The case was discussed with ophthalmology but they did not have sufficient exposure in such a case. We recommend that cases be followed closely following surgery with imaging ever 3–6 months for at least 2 years to monitor for recurrence.

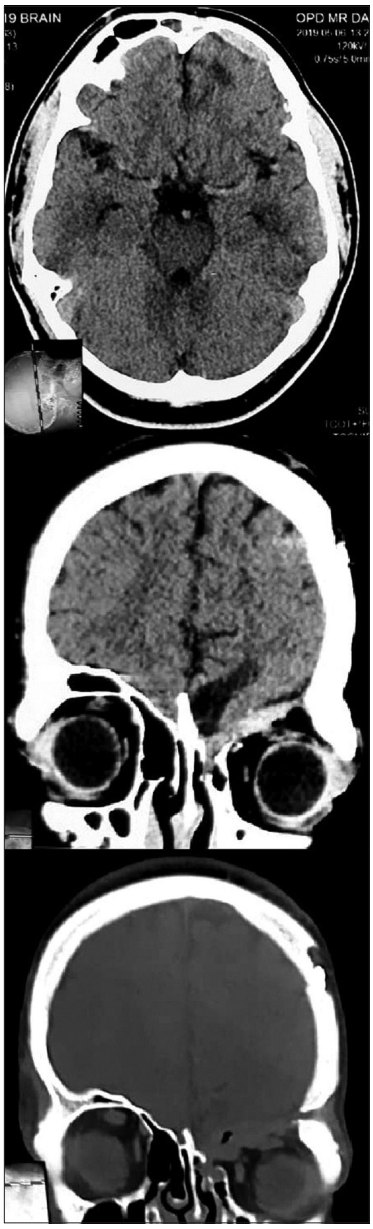

Figure 7: Follow up CT head at two years following surgery showed no evidence of recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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