Rare disease

Juvenile psammomatoid ossifying fibroma: a rare cause of unilateral proptosis

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Summary

This is case of a young girl who presented with unilateral proptosis. Imaging (CT & MRI) revealed an expansile lytic bony tumour involving the left frontal bone and orbital roof. The globe was displaced but uninvolved. Complete surgical resection was done and histopathology revealed psammomatoid bodies diagnostic of juvenile psammomatoid ossifying fibroma (JPOF). JPOF is a rare bone tumour characterised by a predilection for the sino-nasal tract and orbit, a tendency to affect younger patients, a potential for aggressive growth and a high recurrence rate of 30–50%. It warrants a complete surgical resection to avoid recurrence.

BACKGROUND

Juvenile psammomatoid ossifying fibroma (JPOF) is a rare fibro-osseous lesion involving the craniofacial skeleton.¹ It preferentially involves the periorbital, frontal or ethmoid bones and usually presents as nasal obstruction or proptosis. It is locally aggressive and has a high rate of recurrence unlike other fibro-osseous lesions of this region.

Radiologically, it appears as an expansile radiolucent lesion with thick enhancing rim, an appearance which is often non-specific and can occur in other benign tumours of this location.² On histopathology, it is well circumscribed and shows bone trabeculae lined with osteoblasts distributed in vascularised fibrous stroma. The pathognomonic feature, however, is the predominance of psammomatoid bodies.³ Surgical excision is the treatment of choice which must be complete in order to avoid recurrence.

Awareness of the clinical, radiological and pathological appearances of this entity along with a high index of suspicion aids in timely diagnosis and management.

CASE PRESENTATION

A 15-year-old girl presented to the ophthalmic outpatient department with progressive protrusion of left eye for past 6 months. On examination, there was infero-lateral displacement of the globe with normal movements and a vision was 6/6 in both eyes.

INVESTIGATIONS

A CT scan of orbit was done, which revealed an expansile lytic bony lesion measuring 3×3×2.8 cm involving the left frontal bone with extension onto the orbital roof (figure 1). The lesion was predominantly radiolucent with a thick osseous rim and some soft tissue component which enhanced moderately on contrast administration. On MRI, this lesion was hypointense to isointense on T1W and heterogeneously hyperintense on T2W imaging (figure 2A).

It revealed irregular rim enhancement on postcontrast images (figure 2B). Globe was displaced inferiorly; however, the extraocular muscles, optic nerve and chiasma were uninvolved.

DIFFERENTIAL DIAGNOSIS

Based on imaging features, differential diagnoses of benign fibro-osseous lesion, aneurysmal bone cyst (ABC), monostotic fibrous dysplasia and giant cell granuloma were considered.

TREATMENT

The patient underwent frontal craniotomy with extradural tumour removal via left subfrontal approach and postoperative CT showed total removal of the mass.

OUTCOME AND FOLLOW-UP

Histological sections showed a tumour well demarcated from surrounding native bone, composed of fibroblastic stroma containing small ossicles (psammomatoid bodies) (figure 3). The connective tissue stroma was densely cellular with uniform, stellate or plump spindle shaped cells and sparse collagen fibres. The ossicles had thick irregular osteoid rim and coalesced at the periphery to form irregular thin bony trabecular (figure 4). Mitotic activity was rare. A shell of normal bone was present at the periphery showing endosteal osteoclastic activity. Histological features were diagnostic of JPOF.

Patient had an uneventful postoperative period and there was no recurrence at 1-year follow-up.

DISCUSSION

The term 'juvenile ossifying fibromas (JOFs)' is used to describe two distinct clinicopathological entities, the trabecular JOF (TJOF) and psammomatoid JOF (JPOF). The psammomatoid type affects predominantly the

Figure 1 Coronal CT image (bone window) showing an expansile lytic lesion involving left frontal bone and orbital roof.

Figure 2 (A) Coronal T1W MRI image reveals a soft tissue mass isointense to muscle with a peripheral hypointense rim. (B) Coronal post-contrast T1W image showing enhancement of the peripheral bony rim.

Figure 3 Microphotograph at low magnification (10 $\times$) reveals uniformly distributed small ossicles (psammomatoid bodies) in a cellular connective tissue stroma.

Figure 4 At higher magnification, the ossicles show irregular osteoid rim. The intervening stroma consists of plump fibroblasts.

extragnathic craniofacial bones, particularly periorbital, frontal and ethmoid and occurs in 16–33-year age group. On the contrary, the trabecular type involves gnathic bones (maxilla and mandible) at occurs in 8–12-year age group. There is no sex predilection in either group.¹

JPOF clinically manifests as nasal obstruction, proptosis, visual complaints (including blindness), restricted ocular movement or papilledema. On CT scan, it appears as an expansile, usually radiolucent and multilocular lesion with a surrounding thick shell of bone density.¹ On MRI,

a cystic mass with a thick hypointense rim suggestive of osteoid matrix calcification is seen. A JPOF is hypointense to muscle on T1W and T2W sequences. A peripheral rim enhancement is invariably seen on gadolinium-enhanced scans. The enhancement of bony wall suggests that it is active tumour tissue and not reactive sclerosis.²

On imaging, it need to be differentiated from focal fibrous dysplasia (FD), ABC, osteoblastoma, giant cell reparative granuloma or occasionally a giant cell tumour or osteosarcoma if the lesion is aggressive.¹

JPOF usually is punched out lesion with well-defined borders as against FD which has ground glass appearance with indistinct walls. JPOF has a predominantly solid internal structure whereas ABC is cystic with fluid–fluid levels. Imaging features alone may not be diagnostic; nonetheless, in an appropriate clinical setting, they are invaluable for diagnosis.¹

On gross examination, tumour is usually yellowish, white and gritty. On light microscopic examination, it is composed of uniform small ossicles resembling psammoma bodies embedded in a cellular stroma. The ossicles have thick irregular osteoid rim and at the periphery, these ossicles coalesce to form irregular trabeculae.

The psammoma bodies are basophilic and bear superficial resemblance to dental cementum, but usually have an osteoid rim. Because of this superficial resemblance, the lesion has occasionally been mislabelled as cemento-ossifying fibroma. On histology, psammomatoid JOF needs to be differentiated from trabecular JOF and extracranial meningioma with psammoma bodies. JOF is trabecular rather than being psammomatoid, besides having different site and age of presentation. Psammomatoid ossicles of JOF are different from acellular spherical true psammoma

bodies of meningioma. Moreover, meningioma demonstrates EMA positivity which is absent in JOF.⁴

Surgery is the treatment of choice. Complete surgical resection should be the goal of treatment regardless of the technique used, as high recurrence rate up to 30–50% have been reported. If complete resection is not feasible, a close radiological follow-up is recommended as adjuvant therapy such as irradiation has no proven efficacy.

Learning points

- JPOF is a rare locally aggressive fibro-osseous lesion involving the craniofacial skeleton.
- An aggressive surgical treatment is mandatory to avoid recurrence.
- Clinicopathological correlation particularly with radiographic features is essential in establishing a specific diagnosis.

Competing interests None.

Patient consent Obtained.

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