

Hybrid psammomatoid ossifying fibroma & aneurysmal bone cyst of frontal sinus

Munir J Nasser, Mohamed Shawarbi

Abstract: Psammomatoid ossifying fibroma is a rare slowly progressive tumour of the extragnathic craniofacial bones, with a tendency toward locally aggressive behaviour. The pathognomonic histologic feature is the presence of spherical ossicles which are similar to psammoma bodies. A fibro-osseous lesion composed of psammomatoid ossifying fibroma and aneurysmal bone cyst of the frontal sinus in a 12-year-old boy is reported, followed by a literature review. To our knowledge, this is the second case of a hybrid fibro-osseous lesion composed of ossifying fibroma and aneurysmal bone cyst to be reported in the literature.

Key words: Frontal sinus and psammomatoid ossifying fibroma. (p97-99)

Introduction

Psammomatoid ossifying fibroma, also known as juvenile ossifying fibroma, is a rare tumour of the extragnathic craniofacial bones, particularly the periorbital, frontal, and ethmoid bones. It is a slowly progressive lesion with a tendency toward locally aggressive behaviour, including invasion and destruction of surrounding tissue, bony erosion and recurrence after surgical excision. The pathognomonic histologic feature is the presence of spherical ossicles which are similar to psammoma bodies.² Herein we report a psammomatoid ossifying fibroma of the frontal sinus in a 12-year-old boy, associated with an aneurysmal bone cyst. Although aneurysmal bone cyst can occur as a secondary change in association with a number of benign and malignant bone lesions, to our knowledge, this is the second case of a “hybrid” fibro-osseous lesion composed of ossifying fibroma and aneurysmal bone cyst to be reported in the literature.

Case Report

A 12-year-old boy presented with progressive left eye proptosis for the last three years. The condition was

diagnosed initially by ENT surgeon as a mucocele of frontal sinus but after endoscopic biopsy of the lesion the pathology result was suggestive of a meningioma. The patient was referred to our neurosurgical care. Computerised tomography (CT) (Fig. 1) and magnetic resonance imaging (MRI) (Fig. 2) of the skull revealed a huge cystic mass involving left frontal sinus with expansion of the lesion into the ethmoid and orbital cavities. The patient was submitted to left frontal craniotomy. The lesion was partly cystic and completely extradural. The anterior wall of the frontal sinus and the supraorbital part was destroyed by the tumour (Fig. 3). The tumour was excised completely (Fig. 4) and reconstruction of the supraorbital area performed by acrylic cement that was fixed in place with screws. The patient was discharged home few days later without complications.

The material sent for histopathological study consisted of multiple, grayish-white and grayish-brown tissue fragments and bone measuring 4.5 × 4 × 1.5 cm in aggregate. Microscopic examination revealed proliferated oval to spindly cells that often formed small whorls centred by calcified osteoid masses (ossicles) reminiscent of psammoma bodies (Fig. 5). The lesion was seen focally infiltrating bone. However, there was no significant nuclear pleomorphism and mitoses could not be demonstrated. Immunohistochemically, the lesional cells showed strong and diffuse reactivity for vimentin (Fig. 6). There was no immunoreactivity for epithelial membrane antigen (EMA) (Fig. 7). The lesion was interspersed by large blood vascular spaces in areas. Osteoclastic giant cells and osteoid could be focally demonstrated in the lining of these spaces establishing the presence of an associated aneurysmal bone cyst (Fig. 8). The final diagnosis was “psammomatoid juvenile ossifying fibroma and aneurysmal bone cyst”.

King Fahd Hospital of the University
Al Khobar
Saudi Arabia

Correspondence:

Dr. Munir J Nasser
Department of Neurosurgery
King Fahd Hospital of the University
King Faisal University
PO Box 40010
Al Khobar 31952
Saudi Arabia
Email: munir1nasser@yahoo.com



Figure 1 - Preoperative CT sagittal and axial views.

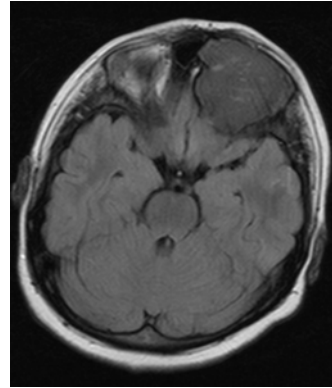


Figure 2 - Preoperative MRI T1-WI axial view.



Figure 3 ← 3DCT scan of skull showing destroyed left supraorbital bone.



Figure 4 → Postoperative CT.

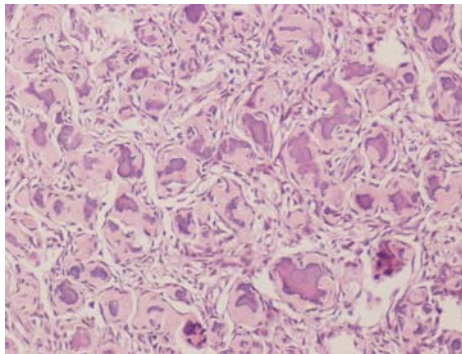


Figure 5 - Microscopic view shows calcified ossicles resembling psammoma bodies.

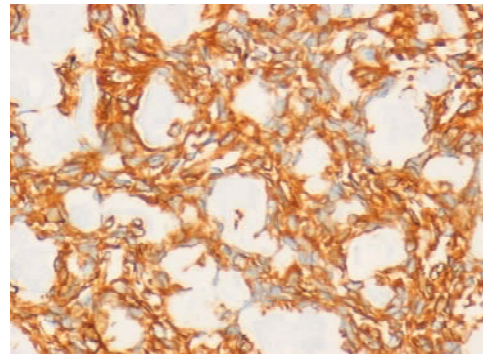


Figure 6 - Immunohistochemically cells showed strong and diffuse reactivity for vimentin.

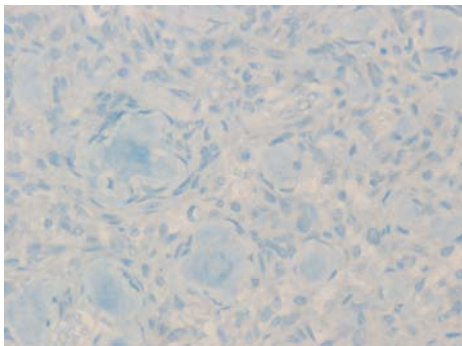


Figure 7 - No immunoreactivity for EMA.

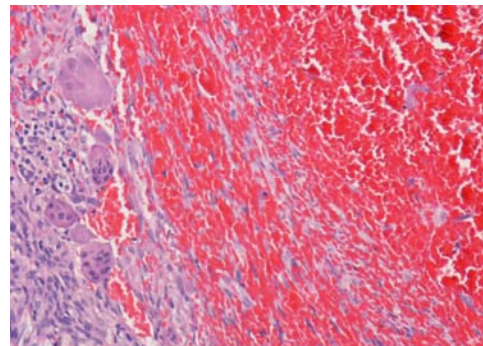


Figure 8 - Large vascular spaces.

Discussion

Psammomatoid ossifying fibroma was initially described by Gogl in 1949 as “psammomatoid fibroma of the nose and paranasal sinuses”.³ In 1985, Margo et al, described psammomatoid ossifying fibroma as a distinctive solitary fibro-osseous lesion of young people that affects the orbit and shows characteristic histologic features.⁷ Psammomatoid ossifying fibroma has also been reported under the designations “psammous desmo-osteoblastoma” by Makek⁽⁶⁾, “juvenile active ossifying fibroma” by Johnson et al,⁽⁵⁾ and “juvenile ossifying fibroma” with psammoma-like ossicles by Slootweg et al.⁸ It probably arises from overproduction of the myxofibrous cellular stroma normally involved in the development of the septa in the paranasal sinuses as they enlarge and pneumatize. These stromal cells secrete hyaline material that ossifies and connective tissue mucin that initiates the cystic areas. The majority of cases have been reported in children and young adults between 5 - 25 years of age with slight male predominance (1.2:1). The lesion is particularly common in the periorbital, frontal and ethmoid bones. Our patient was a 12-year-old boy and the lesion was located in the frontal sinus. Clinically, psammomatoid ossifying fibroma presents with proptosis, visual disturbances, blindness, ptosis, disturbances in ocular mobility, papilloedema, recurrent headaches and nasal obstruction. Radiographic examination reveals an expansile well-circumscribed radiolucent or mixed radiolucent/radiopaque lesion surrounded by a thick bony wall.⁴ The present case presented with proptosis. Computerised tomography and MRI of the skull revealed a huge cystic mass involving left frontal sinus with expansion of the lesion into the ethmoid and orbital cavities. The cystic nature of the present lesion which was also verified intraoperatively may be attributed to the presence of an associated aneurysmal bone cyst. A hybrid fibro-osseous lesion composed of psammomatoid ossifying fibroma and aneurysmal bone cyst have been

previously reported.¹ The most characteristic histologic feature of psammomatoid ossifying fibroma is the presence of numerous small, round ossicles or “psammomatoid” bodies that are embedded in a cellular fibrous stroma. The ossicles are mineralized collagenous foci that vary from small, smoothly contoured round-to-oval patterns to larger, irregularly shaped patterns, with concentric layering similar to that of psammoma bodies. A prominent marginal osteoid rim surrounds the ossicles. The main histologic differential diagnosis is a primary sinonasal meningioma. The completely extradural location and the partly cystic nature of the present lesion, the young age of the patient and the EMA negativity of the tumour cells favour the diagnosis of psammomatoid ossifying fibroma over meningioma.

References

1. Blayney AW, El Tayeb AA: The 'hybrid' fibro-osseous lesion. *J Laryngol Otol* 1986, 100(3): 291-302
2. El-Mofty S: Psammomatoid and trabecular juvenile ossifying fibroma of the craniofacial skeleton: two distinct clinicopathological entities. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2002, 93: 296-304
3. Gogl H: Das Psammo-osteoid-fibrom der nase und ihrer nebenhohlen. *Monatsschr Ohrenheilkd Laryngo-Rhinol* 1949, 83: 1-10
4. Han MH, Chang KH, Lee CH, Seo JW, Han MC, Kim CW: Sinonasal psammomatoid ossifying fibromas: CT and MRI manifestations. *Am J Neuroradiol* 1991, 12(1): 25-30
5. Johnson LC, Yousefi M, Vinh TN, Heffner DK, Hyams VJ, Hartman KS: Juvenile active ossifying fibroma. Its nature, dynamics and origin. *Acta Otolaryngol* 1991, 488(Suppl): 1-40
6. Makek MS: So called “fibro-osseous lesions” of tumorous origin. Biology confronts terminology. *J Craniomaxillofac Surg* 1987, 15(3): 154-167
7. Margo CE, Ragsdale BD, Perman KI, Zimmerman LE, Sweet DE: Psammomatoid (juvenile) ossifying fibroma of the orbit. *Ophthalmol* 1985, 92(1): 150-159
8. Slootweg PJ, Panders AK, Koopmans R, Nikkels PG: Juvenile ossifying fibroma. An analysis of 33 cases with emphasis on histopathological aspects. *J Oral Pathol Med* 1994, 23(9): 385-388