CASE REPORT

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Psammomatoid ossifying fibroma*

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SUMMARY

Psammomatoid ossifying fibroma (POF) represents a unique subtype of fibro-osseous lesions. We describe a case of POF involving the orbit and the sinonasal tract, in a 13-year-old white female. Diagnosis depends on the histological, radiological and clinical features. Complete excision by endoscopic nasal surgery was the treatment of choice. Five years later the patient was free from symptoms and tumour recurrence. Differential diagnoses are discussed.

Key words: orbital tumour, bone tumor, psammomatoid, ossifying fibroma, fibro-osseous lesion

INTRODUCTION

Psammomatoid ossifying fibromas (POF) represents a rare subtype of fibro-osseous lesions. It is a slowly progressive lesion with the potential to create sizable defects in the orbit and the sinonasal tract. Clinical manifestations include proptosis, nasal obstruction, headache and facial swelling.

In this case report, we describe a case of POF in a 13-year-old white female.

CASE REPORT

The patient was referred to the ENT specialist consultation for evaluation of a right side proptosis of one year duration. Other symptoms included nasal obstruction and facial swelling. A mucosal appearance tumour was noted obstructing the right nasal fossa and arising from the lateral wall.

Computed tomography presented a well demarcated expansive lesion extending into the ethmoidal, maxillary and sphenoid right sinus and invading the right nasal fossa. Exophtalmus and orbital compression by the tumour were seen in CT scans (Figure 1).

Excision by endoscopic nasal surgery was the treatment of choice. The wall of the mass, composed of a mucous membrane and bone trabeculae, was perforated and removed, draining a viscous chestnut coloured liquid.

The histological appearance was characterized by the presence of small mineralised bodies admixed with a dense cellular stroma. Bony trabeculae surrounded by osteoblasts and multinucleate giant cells were found (Figures 2 and 3). Atypia and mitotic activity could not be observed. Due to the infrequent features, analysed material was sent to the Memorial Sloan-Kettering Cancer Center (New York), where Doctor Juan Rosai confirmed the histological diagnosis.

Five years after complete surgical removal the patient was free from symptoms and tumour recurrence.

DISCUSSION

The concept of maxillofacial fibro-osseous lesions includes three major entities: fibrous dysplasia, ossifying fibroma and periapical cemental dysplasia (Waldron, 1985; Kramer et al., 1992; Slootweg, 1996). Diagnosis depends on the histological, radiological and clinical features.

Histologically, fibrous dysplasia shows evenly distributed islands of woven bone that fuse with surrounding bone. However, especially in the jaws and in older patients, fibrous dysplasia may contain lamellar bone. Radiographs typically show absence of a sharp line of demarcation. Fibrous dysplasia usually is a self-limiting disease. Patients may require surgical intervention for cosmetic reasons, but complete removal of the lesion in most instances is impossible and also unnecessary (Slootweg et al., 1990; Waldron, 1993).

In contrast, ossifying fibroma has a tendency toward locally aggressive behavior and has to be removed completely. It is characterized by a fibrous tissue and mineralized material that, in contrast with fibrous dysplasia, does not fuse with its surroundings. Thus, the lesion is demarcated or incapsulated. On the opposite, osteosarcomas perforate the cortical plate and spread into adjacent soft tissues. Ossifying fibroma shows variable amounts of woven and lamellar bone trabeculae and osteoblasts may rim trabeculae (Eversole et al., 1985; Slootweg et al., 1990). Recently, a rare case of ossifying fibroma of the middle turbinate has been described (Caylakli et al., personal communication).

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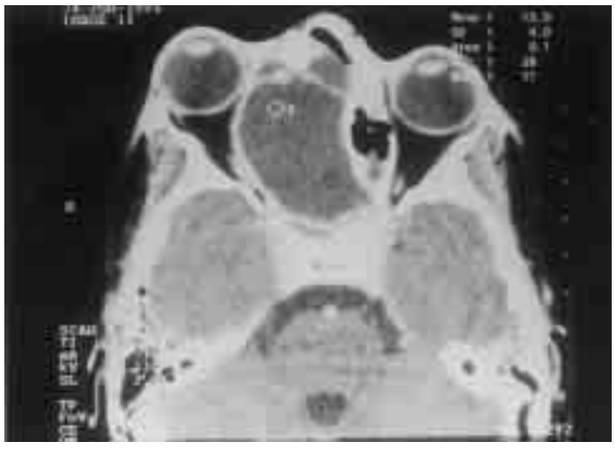
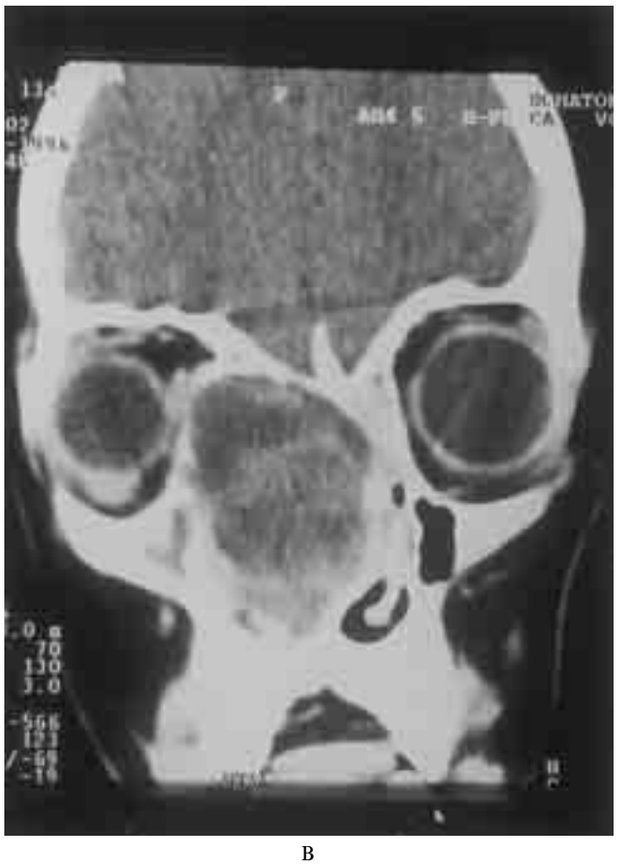


A



B

Figure 1. CT scan showing psammomatoid ossifying fibroma in the right ethmoid and sphenoid (A) and invading right nasal fossa and extending into the ethmoidal, maxillary sinus, causing orbital compression (B).

Psammomatoid is a subtype of ossifying fibroma and has distinctive histomorphological and clinical features. The histological appearance is characterized by the presence of small rounded mineralised (psammmomatoid) bodies admixed with stroma that varies from being loose and fibroblastic to intensely cellular without intervening collagen.

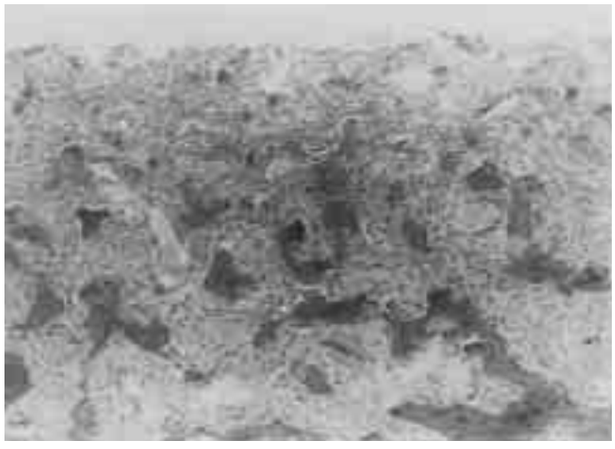


Figure 2. Psammomatoid ossifying fibroma. Typical pattern of anastomosing trabeculae of woven bone separeted by cellular fibrovascular strome.

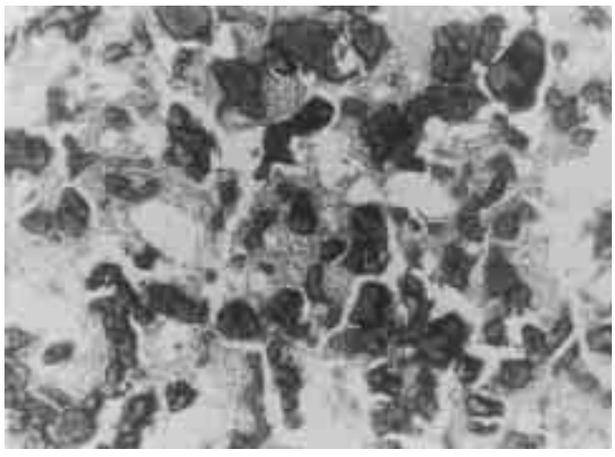


Figure 3. Psammomatoid ossifying fibroma. Anastomosing trabeculae of bone with predominant psammomatoid pattern of matrix mineralization.

It is typically a childhood disease, although the lesion can occur in adults. Males and females are equally affected. The most common symptoms and signs are proptosis, facial swelling, nasal obstruction, headaches and sinusitis (Margo et al., 1985; Wenig et al., 1995). All paranasal sinuses, particularly the ethmoid and maxillary sinuses, and lateral nasal wall can be involved (Wenig et al., 1995; Danielides et al., 2002). Radiographic studies show the presence of an osseous or soft tissue mass varying in appearance from well demarcated without invasion or erosion to invasive with bone erosion and intracranial extension (Wenig et al., 1995). In this case, the lesion was completely excised and has not recurred.

Finally, periapical cemental dysplasia is confined to the jaws, near the tooth apices. Its histologic features are similar to

those of ossifying fibroma but without a sharply defined margin (Kramer et al., 1992). The removal of the lesion in unnecessary because it does not cause any clinical symptoms (Slootweg, 1996).

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