PSAMMOMATOID OSSIFYING FIBROMA OF THE ETHMOID

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ABSTRACT

We present a 16 year-old girl who suffered since one year of a painless slow growing mass on the left medial orbital rim causing globe displacement. CT- scan and MRI of the orbit with T1 and T2 weighted images showed the presence of a large mucocoele in the frontal sinus. This occured secondary to the obstruction of the sinonasal tract by a bony tumour. Histopathology showed a lesion consisting of fibrous tissue and ossicles or psammomatoid bodies.

The diagnosis of a psammomatoid ossifying fibroma (POF) was made.

RÉSUMÉ

Présentation d'une patiente de 16 ans souffrant d'une masse orbitaire non douloreuse à évolution lente. Le scanner et l'IRM de l'orbite montraient la présence d'une tumeur à composante osseuse dans le sinus ethmoidal avec déplacement du globe. L'obstruction du tractus sinonasal a provoqué un grand mucocèle du sinus frontal. L'histologie est caractérisée par une lésion fibro-osseuse avec des structures qui ressemblent à des corps psammomateux. Le diagnostic de fibrome ossifiant psammomatoide a été posé.

SAMENVATTING

Een 16-jarig meisje consulteerde omwille van een pijnloze traag groeiende massa ter hoogte van de mediale rand van de linker orbita. Er was een duidelijke laterale oogbolverplaatsing. De CT-scan en MRI van de orbita met T1 en T2 gewogen beelden toonden de aanwezigheid van een tumor met botcomponenten in de ethmoïdale sinus. Door obstructie van de sinonasale tractus ontstond een grote mucocoele in de frontale sinus. De tumor werd verwijderd via externe weg. Microscopisch onderzoek toonde een letsel bestaand uit fibreus stroma met ossikels of ″psammomatoid bodies″. De diagnose van psammomatoid ossificerend fibroom (POF) werd gesteld.

KEY-WORDS

psammomatoid ossifying fibroma, psammomatoid bodies, ossifying fibroma, ethmoidal tumor, orbital tumor

MOTS-CLÉS

Fibrome ossifiant psammomatoide, corps psammomateux, fibrome ossifiant, tumeur ethmoidale, tumeur orbitaire

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INTRODUCTION

Psammomatoid ossifying fibroma (POF) is an uncommon variant of ossifying fibroma that typically occurs in the sinonasal tract (7).
It can cause cosmetic disfigurement and has a tendency to recur. Therefore regular follow-up is necessary after surgery (6). The histologic picture is that of a benign well demarcated fibro-osseous lesion with the presence of ossicles or psammomatoid bodies (3).

CASE REPORT

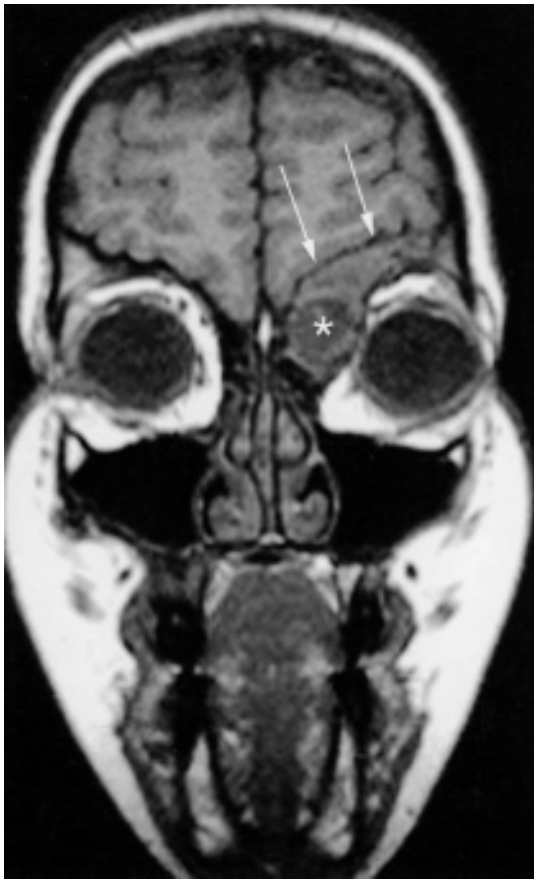
A 16-year old girl developed a slowly progressive lateral displacement of the left globe (fig.1). She had no visual loss, diplopia or pain. On examination the motility and the anterior segment were normal and there were no choroidal folds. There was no afferent pupillary defect.
The patient was examined at the Department of Paediatrics and Head and Neck Surgery.
CT and MRI of the orbit with T1 and T2 weighted images were performed.
A T1-weighted image showed an expanded left frontal sinus secondary to the obstruction by a tumour in the ethmoidal cells (fig.2A/2B). The hyperintense signal content of the left frontal sinus compared to the signal of the ocular vitreous was indicative for a high protein content (mucocoele formation).
After administration of contrast, enhancement was lacking in the lesion, but was noted in the periphery of the lesion (fig.3A/3B).
T2-weighted image revealed absence of signal (signal void) in the tumour, suggesting a bony character of the lesion (fig.4A/4B).
A 3mm thick axial CT image confirmed the bony features of the lesion. The lamina papyracea was displaced towards the left orbit (fig.5).
A careful complete excision was performed by an external approach at the Department of Head and Neck Surgery.
Microscopy revealed a benign, well demarcated fibro-osseous lesion,composed of cellular stroma (non-osseous component) with a fascicular to storiform growth pattern, composed of polyhedral to spindle shaped cells with prominent basophilic nuclei and inapparent cytoplasmic borders (fig.6A/6B).

Fig. 1. a 16 year-old girl with lateral displacement of the left globe

Mitotic figures could be seen.The most distinctive component was the presence of mineralized or calcified ''psammomatoid'' bodies or ossicles. These ossicles varied from small with a round to oval shape to large and irregular shaped.
The patient was examined after the operation. The lateral displacement of the left globe disappeared.

DISCUSSION

The distribution of orbital disease in a 16-year old patient is largely similar to the adult pattern.
Inflammatory conditions are the most common cause of orbital disease (45%), structural abnormalities account for 25% and neoplastic disorders are present in 27% of the cases. Vascular abnormalities are uncommon in this age group (3%) (6). **Psammomatoid ossifying fibroma (POF)** is an example of a neoplastic disorder and its characteristic histologic picture



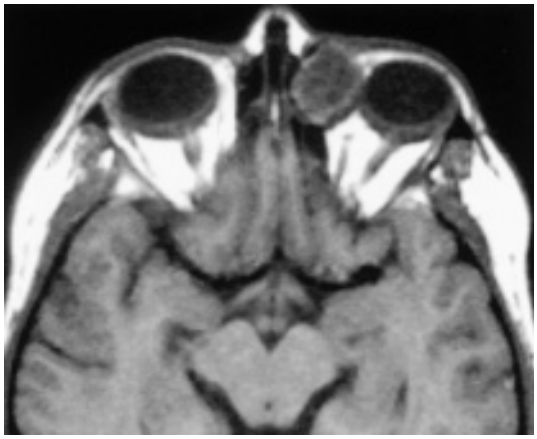


Fig. 2 A and 2B. T1-weighted frontal and coronal MR image of the orbitae. The mass has a low to intermediate signal intensity and is surrounded by a slightly hyperintense margin

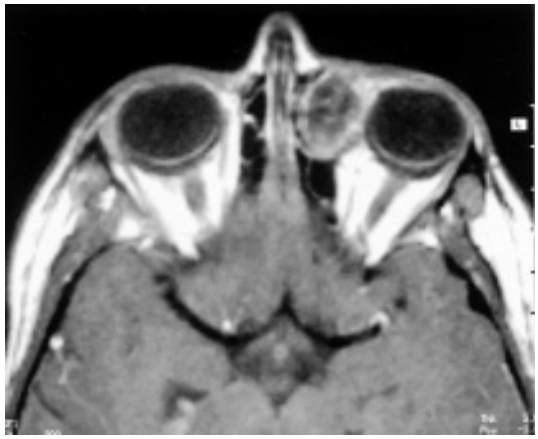


Fig. 3A and 3B. T1-weighted frontal and coronal MR image of the orbitae after gadolinium contrast administration. The margin of the mass shows intense enhancement which corresponds to inflammatory mucosa of the obstructive sinusitis. The osseous delineation is not visible.

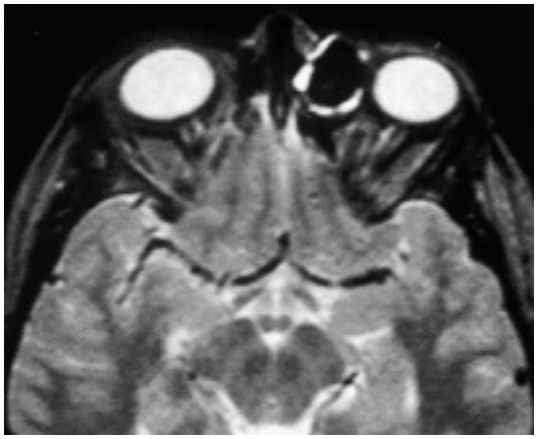


Figure 4A and 4B: T2-weighted frontal and coronal MR image of the orbitae. The lesion is visible as a black area, surrounded by a hyperintense margin, reflecting obstructive sinusitis. The left globe is displaced laterally.

Fig. 5. CT of the orbitae. An inhomogenously calcified mass displaces the lamina papyracea of the left orbit. There is secondary sinusitis of the anterior ethmoidal sinuses. There is no sclerotic delineation around the lesion.

helps to differentiate it from other bone tumours. The word ''psammos'' is derived from the Greek word ''ψαμμοσ'' meaning sand. Psammomatoid bodies, found in psammomatoid ossifying fibromas, are spherules of lamellar bone rimmed by osteoblasts and are reminiscent of psammoma bodies. The onset of psammomatoid ossifying fibroma is often in the mid-teenage years, and the average duration of symptoms before presentation is 4 to 5 years (3). As the lesion can also occur in adults, the use of the term juvenile ossifying fibroma is incorrect (5). Mansour et al reported on a 66 years old woman with POF of the ethmoid sinus who presented with orbital cellulitis (4). POF is a slowly progressive lesion that can create sizable defects in the orbital bones. The treatment is surgical and complete excision is important given the tendency to recur (1,3,5-7). It is a more agressive tumour than typical ossifying fibroma or fibrous dysplasia. Psammomatoid ossifying fibroma may be difficult to distinguish from fibrous dysplasia, osteoma, well-differentiated osteosarcoma and psammomatous meningioma (table).

The clinical features of **fibrous dysplasia** of the orbit are proptosis and downward displacement of the globe. Optic nerve compression is observed in 50% of cases. Malignant transformation is extremely rare (0.5%).CT-scan shows a typical groundglass-appearance. The most common place is the orbital roof. The involved bones

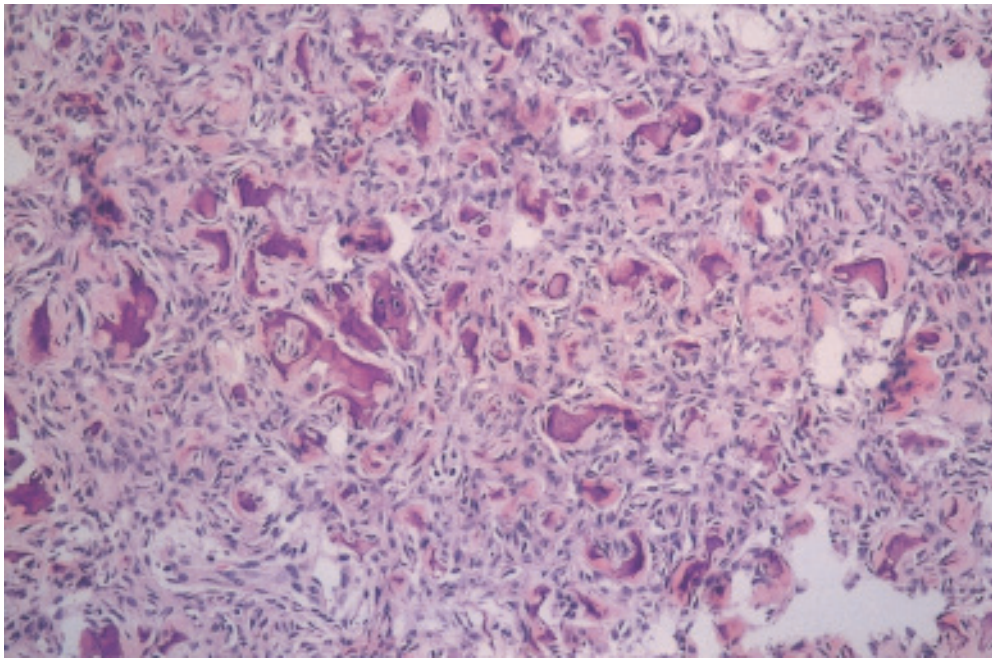


Fig. 6A. Haematoxylin and eosin stain (10x): spindled stromal cells and spherical ossicles with "psammomatoid" aspect

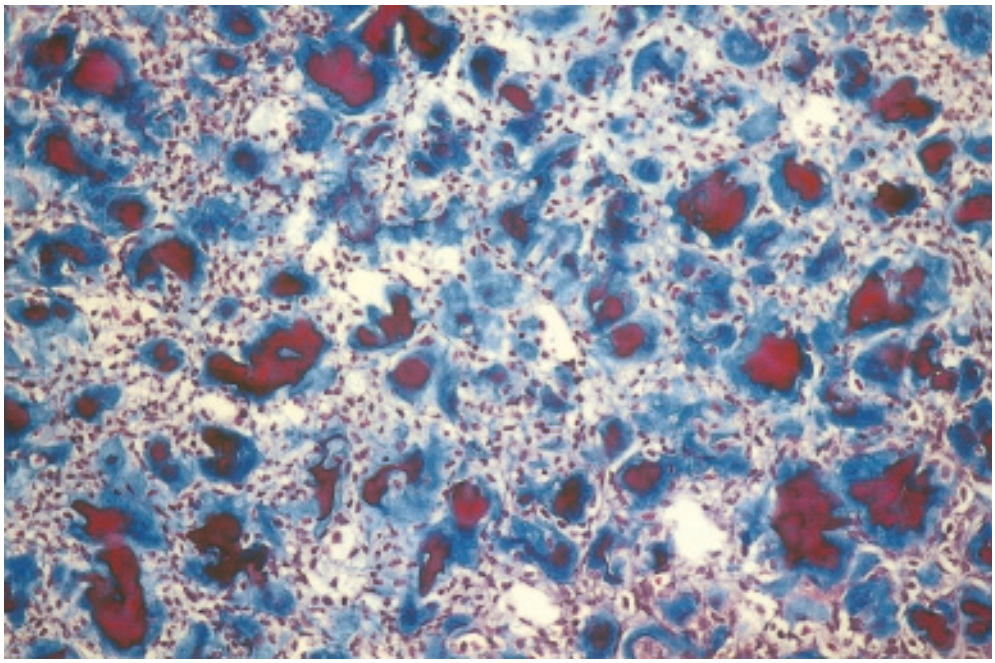


Fig. 6B. Masson's trichrome stain (40x): well demarcated fibro-osseous lesion with cellular stroma and calcified psammomatoid bodies

Table: *Differential diagnosis*

TYPE	CLINICAL FEATURES	RADIOLOGIC FEATURES	HISTOPATHOLOGY	TREATMENT AND PROGNOSIS
Fibrous dysplasia	Progressive and self-limited growth, 1° and 2° decade	Homogenous dense sclerosis or pagetoid aspect (CT)	Bone trabeculae in fibrous background, no osteoblasts	Observation, surgery
Osteoma	Slow growth, 2°-5°decade	Signal void on T1 and T2, moderate enhacement nidus (MRI)	Dense lamellar cortical bone with vascular canals	Observation, surgery
Well differentiated osteosarcoma	Rapid growth with pain, male predominance, 1°-3° decade	Calcifications, lytic and sclerotic changes (CT)	Mimics fibrous dysplasia, but shows nuclear polymorphism	Radical excision and chemotherapy
Psammomatous intracranial meningioma	Slow growth, 6°-7° decade, female predominance	Hyperostosis and lysis, soft tissue tumor enhancement after contrast injection (CT)	Wide range psammoma bodies	Observation, surgery, radiotherapy

are thickened. There is a low signal on T1 with mild enhancement after contrast injection (1) and an heterogeneous signal on T2. Histopathology shows a lesion consisting of bone trabeculae in fibrous background without osteoblasts.

Osteoma of the orbit is the most common bony tumour of the orbit. It is a well differentiated benign tumour consisting of dense lamellar cortical bone with vascular canals (Havers like). The radiologic features of the lesion are a dense mass, often with a mucocoele formation. There is a signal void on T1 and T2-weighted MR images and homogenous very high density of compact bone on CT.

Osteosarcoma of the orbit can occur after irradiation. Survivors of the genetic form of retinoblastoma are at greater risk (7). There is a male predominance. The prognosis is poor (6). On CT a massive destructive lesion is seen with evidence of calcification (1). There is a wide range of histological appearances.

An **extra-optic psammomatous meningioma** is also an important differential diagnosis. There is a wide range of histopathological appearances and psammoma bodies can be present. The survival is exellent. Fine calcifications can be present. There is enhancement in the lesion after contrast injection.

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