Quiz

Correct answer to the quiz. Check your diagnosis

Case report

Juvenile psammomatoid ossifying fibroma

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Juvenile psammomatoid ossifying fibroma (JPOF) is an uncommon benign and locally aggressive tumor. We report an unusual head tumour with extremely rare extensiveness and aggressivness. The patient was 18-year-old female with three-day-lasting headache and repetitive oral bleeding. Computed tomography revealed a large, well-circumscribed, expansile mass occupying ethmoid cells, nasal cavities and ventral part of the sphenoid sinus, with extention into the anterior cranial fossa. Pterional craniotomy was carried out. On one-year follow-up recurrence of the lesion was identified and the second surgery was performed. The lesion is under supervision now, due to incomplete removal.

Key words: ossifying fibroma, psammomatoid juvenile ossifying fibroma, sphenoid sinus, ethmoid sinus, anterior cranial fossa.

Introduction

Juvenile psammomatoid ossifying fibroma (JPOF) is an uncommon benign and locally aggressive tumor. It is a neoplasm composed of the stroma containing small ossicles resembling psammoma bodies [1]. The lesion occurs in the craniofacial bones of young adults, especially in the periorbital frontal and ethmoid ones [1]. JPOF continues to grow when left untreated [1]. Recurrence is usually due to incomplete removal, therefore, it is recommended to resect it completely [2].

The aim of this work is to report the clinical and histologic details of an extensive and locally aggressive JPOF involving the nasal cavity, sphenoid and ethmoid sinus with extention into the anterior cranial fossa.

Case report

An 18-year-old, healthy female, with the history of hyperpituitarism in 2004 and subsequent adenoidectomy in 2006, was consulted in University Clinical Center in Gdansk with three-day-lasting headache and repetitive oral bleeding in June and August 2015.

Computed tomography revealed a large, well-circumscribed, expansile mass occupying ethmoid cells, nasal cavities and ventral part of the sphenoid sinus. Additionally, the lesion extended into the anterior cranial fossa. In subsequent magnetic resonance imaging (MRI) the lesion presented as a polycystic tumor with fluid-fluid levels. On T1-weighted images, the non-cystic areas had intermediate signal intensity

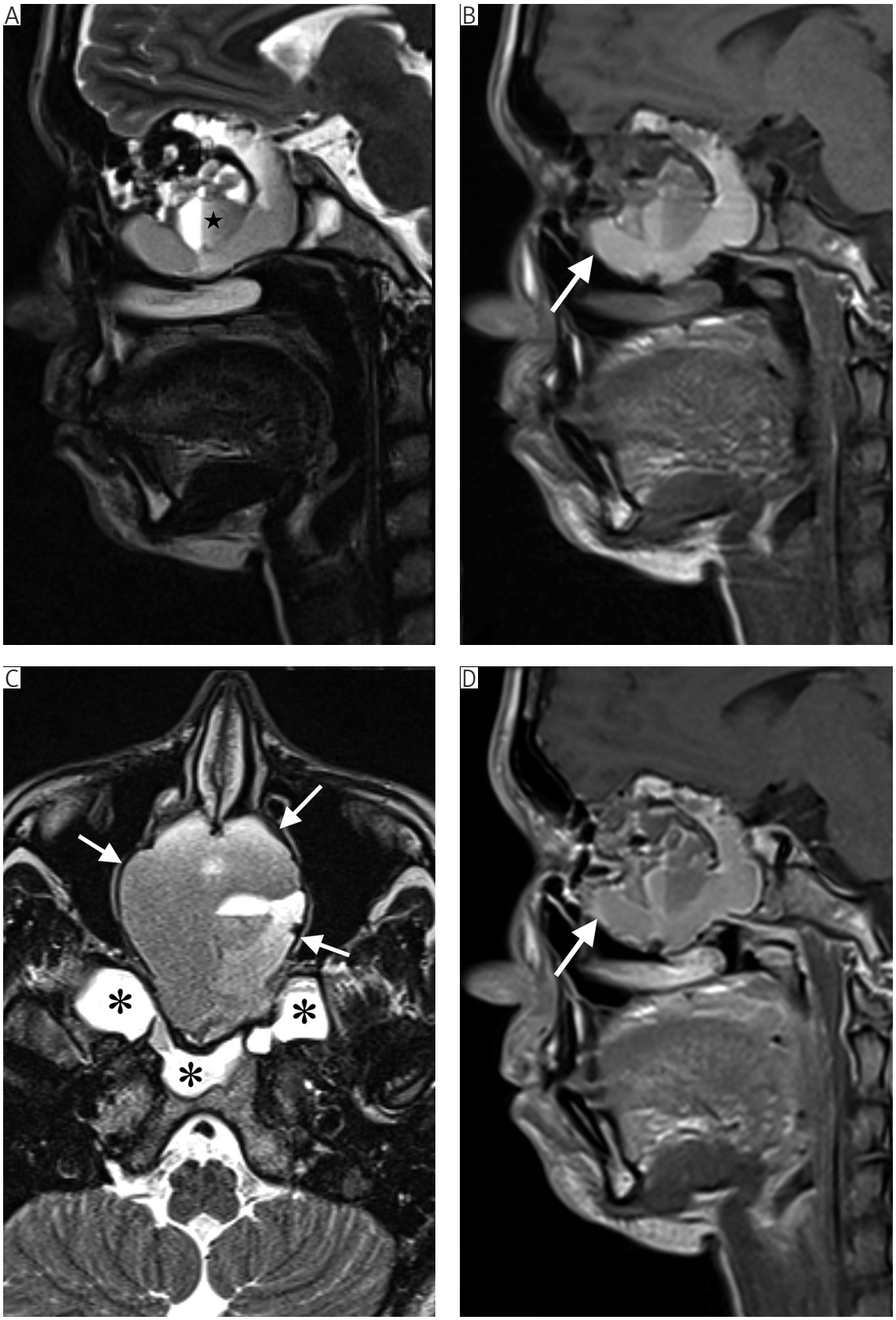

Fig. 1. Initial MRI of the patient. Figure A presents fluid-fluid levels (black star) within the lesion in the T2-weighted image in the sagittal plane. T1-weighted images in turbo spin echo sequence in the sagittal plane without (B) and with contrast enhancement (D), the lesion presents a mild contrast enhancement (white arrow) in non-cystic, peripheralareas. In figure C a low signal intensity boundis visible (white arrows) on the T2-weighted image in the axial plane, which corresponds with the bony rim. Additionally, mucocele in the sphenoid sinus is visible (asterisks)

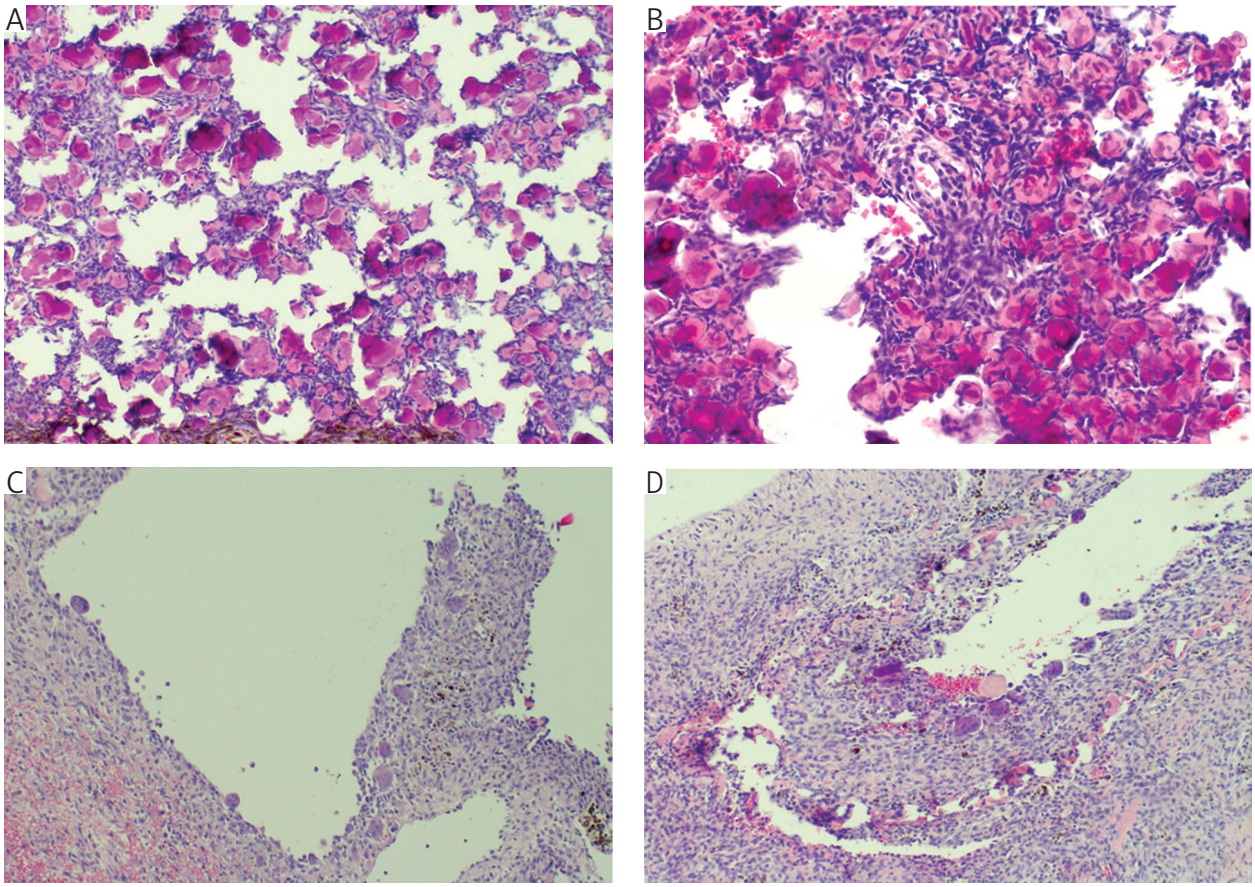

Fig. 2. Microscopic view shows calcified ossicles resemble psammoma bodies and cellular stroma (A – HE, 100×, B – HE, 200×), cystic part with giant cells and haemorrhages (C, D – HE, 100×)

with enhancement after contrast medium administration. The lesion obstructed the sphenoid sinus causing mucocele (Fig. 1). An aneurysmal bone cyst (ABC) was suspected.

Pterional craniotomy was carried out and the tumour was removed incompletely. On microscopic examination the lesion was cellular with bland, monomorphic spindled cells dispersed in the stroma which contained numerous, small, round bony psammoma-like bodies. The stromal cells were negative for EMA, CD34, S100, PR, CK AE1/3. Cystic degeneration with ABC-like structures was observed. These cysts were lined by fibrous wall with giant cells and haemorrhages (Fig. 2). Final diagnosis of JPOF was rendered.

On one-year follow-up recurrence of the lesion was identified (Fig. 3). Therefore, the second surgery was performed, but the resection was incomplete. The lesion did not show any histological signs of malignancy. No further treatment was applied.

Discussion

The term ossifying fibroma (OF) is used to describe a benign, but potentially aggressive, bone-producing fibrous neoplasm of the craniofacial skeleton [3]. The current WHO classification distinguishes three similar histological variants of OF: cemento-ossifying fibroma (COF), juvenile trabecular ossifying fibroma (JTOF) and JPOF [1]. They were believed to have odontogenic origin [1], however, the recent research suggested JPOF and JTOF are nonodontogeniclesion [2].

COF is mainly seen in young adults. The onset of JTOF and JPOF is usually in childhood and adolescence [2, 4]. This is in accordance with our case of an 18-year-old girl.

COF is generally localized in the mandible with predilection to premolar and molar areas. Its growth is slow, with lingual and buccal expansion. Displacement of teeth is observed as the lesion expands [2, 5]. JTOF is mostly seen in the mandible and maxilla, affects predominantly the non-dental areas and grows rapidly [2]. JPOF occurs in paranasal sinuses, particularly in the frontal and ethmoid, but the lesion was also reported in the calvarium and the mandible [6]. It may penetrate the orbit and anterior cranial fossa. The bony expansion is the most common clinical manifestation. It can show various symptoms, e.g. proptosis, nasal obstruction, headaches, facial swelling,

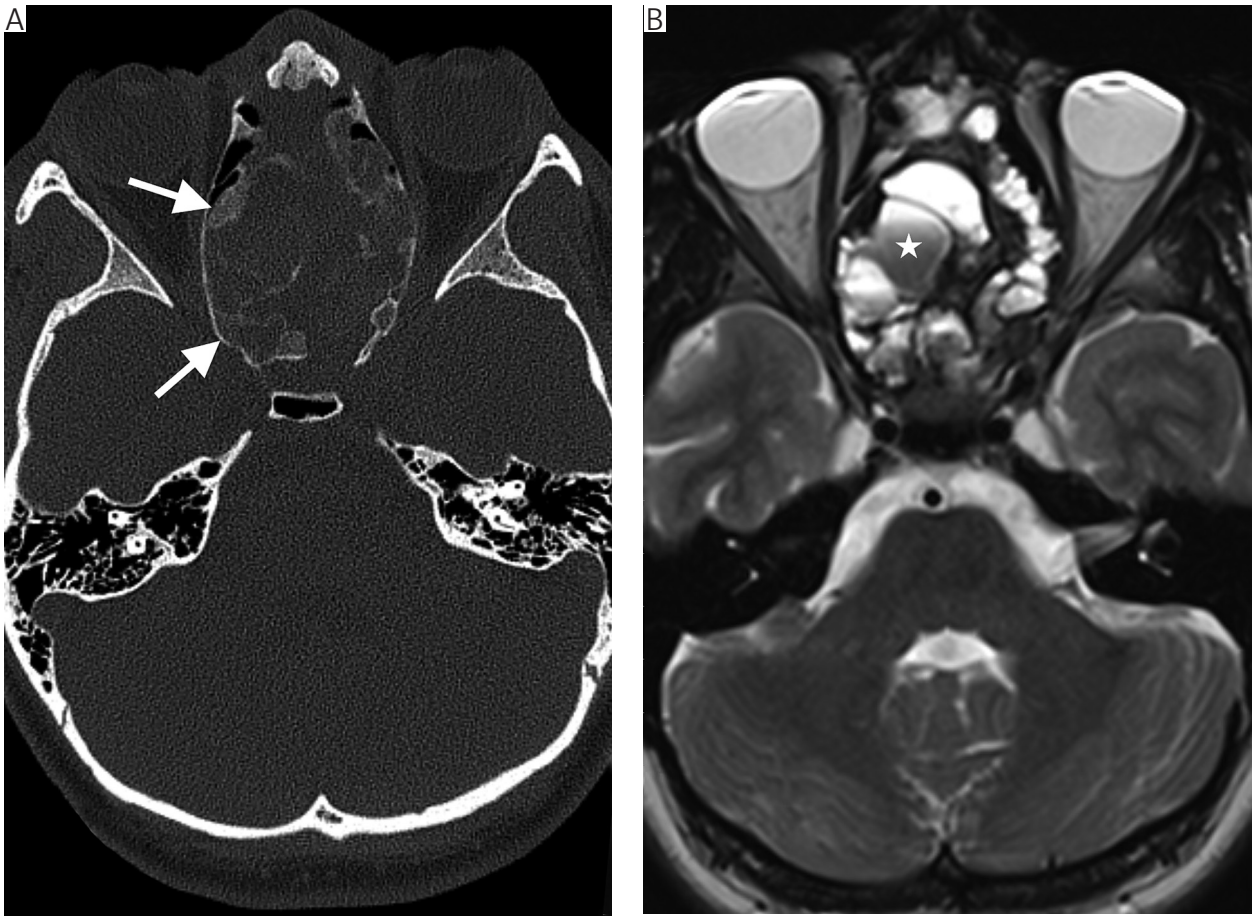

Fig. 3. Follow-Up examinations shows recurrence of the lesion in non-enhanced computed tomography in the hard kernel and bone window (A) and T2-weighted image in the axial plane (B). White arrows indicate bonny parts of the lesion in peripheral areas and the white star is in a cystic part of the lesion with fluid-fluid level

pain and recurrent sinusitis [5]. It may also have an aggressive course [7].Our case developed in the nasal cavity and ethmoid sinus with extension into the anterior cranial fossa and affected also the sphenoid sinus, the site which has not been reported yet.

COF is composed of fibrous tissue that varies from intensely cellular to acellular areas. Examination reveals presence of a cementum-like material, blood vessels, woven and lamellar bone [2, 5]. JTOF comprises of bands of cell-rich osteoid encasing osteocytes and highly cellular fibrous stroma surrounded by slender trabeculae. JPOF is characterized by fibroblastic stroma containing spherical calcifications resembling psammoma bodies. The stroma varies from being hipercellular (usually) to fibroblastic and loose in texture [1, 2]. Cystic degeneration and aneurysmal bone cyst-like formation are commonly observed [8, 9]. These cysts are composed of thin fibrous wall and usually comprise osteoclast giant cells and haemorrhages [2], similarly to our case.

On radiographs, COF is well demarcated with variable radioopacity. Radiological imaging of JTOF and JPOF shows a well circumscribed lesion, its area may be radiolucent or radiopaque depending on the soft and hard tissue components [2, 9].

Apart from other fibro-osseous lesions, JPOF has to be distinguished from fibrous dysplasia, osteoblastoma, osteosarcoma, primary aneurysmal bone cyst. Intraosseous cavernous hemangioma and eosinophilic granuloma should also be taken into account due to radiologic and histologic similarities [10, 11]. JPOF may be misdiagnosed as extracranial menigioma with psammoma bodies, which is EMA positive, in contrast to JPOF [12]. JPOF is negative for S100 and CD34 [12], Our case is EMA, S100 and CD 34 negative.

A successful treatment of these craniofacial tumors is frequently problematic [13]. Recurrence is usually due to tumor persistence after incomplete removal and its rate ranges from 30 to 56% [8]. Enucleation and curettage may be succesful treatment for small JPOF, the larger and more aggressive tumours require resection with 5 mm margin [12]. Due to the tumor location, it would have to be removed down to the level of unchanged bone with preservation of contiguous vital structures as far as possible [12]. The location and extent of the tumor

determine surgical approach(intracranial or a combined subfrontal and nasal) [14]. Endoscopic resection may be successful in some endonasal cases [15]. It is believed the lesion is radio-resistant, therefore, radiotherapy is contraindicated [16]. Some data proved the alpha-interferon therapy in combination with surgical curettage is an alternative treatment [13]. Clinical and radiological follow-up is requisite for JPOF. Malignant transformation was not reported in the literature [8]. In conclusion, the prognosis remains unfavorable in our case due to tumour location and its aggressive potential.

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