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Case Report

Psammomatoid ossifying fibroma in the extraconal space: a case report

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Abstract: A 24-year-old patient with left eye proptosis and intermittent pain for 5 months was admitted to our hospital. Physical examination revealed neither extra ocular muscle limitations nor visual field defects. Magnetic resonance imaging (MRI) revealed a multicystic mass in the left extraconal space compressing the superior oblique muscle and adjacent frontal lobe. Layered hemorrhage was observed within the lesion in the 1-month follow-up MRI. Dynamic contrast enhanced imaging showed mild increased perfusion of the surrounding peripheral portion. Magnetic resonance spectroscopy showed an increased lactate/lipid peak of 1.3 ppm. Combined open and endonasal surgery was performed, and the final diagnosis was psammomatoid ossifying fibroma. The tumor was positive for vimentin, and negative for smooth muscle actin, S100 and epithelial membrane antigen. Despite its rarity, psammomatoid ossifying fibroma should be considered when multicystic lesions with peripheral enhancement near the orbit exhibit progressive inner hemorrhage.

Keywords: Juvenile psammomatoid ossifying fibroma, vimentin, magnetic resonance imaging, computed tomography, skull base

Introduction

Ossifying fibroma (OF) usually affects the craniofacial area during the first two decades [1, 2]. OF is a fibrous-osseous tumor that is histologically characterized by fibrous cellular stroma with a mixture of mineralization and ossification foci [3]. OF is benign, and because of its local destructive pattern, variable imaging appearances, and rapid growth rate, accurate diagnosis can be challenging [4]. There are three kinds of OF, namely, juvenile psammomatoid ossifying fibroma (JPOF), cement-ossifying fibroma (COF), and juvenile trabecular ossifying fibroma (JTOF) [3]. Herein, we reported a case of JPOF with an evolving appearance on follow-up imaging.

Case report

A 24-year-old patient was admitted to our hospital due to left eye proptosis and mild intermittent pain for 5 months. Physical examination revealed no extra ocular muscle limitation, visual field defects, or diplopia.

The patient underwent magnetic resonance imaging (MRI), which revealed a 28 × 22 × 24 mm T1 iso-signal and T2 hyper-signal-intensity lesion with a multicystic appearance compressing the superior oblique muscle and frontal lobe in the left orbit (**Figure 1A**). T2-weighted imaging revealed layered blood-fluid levels (**Figure 1B**). Contrast-enhanced T1-weighted image revealed peripheral rim enhancement of the lesion (**Figure 1C**) without diffusion restriction (**Figure 1D**). Layered hemorrhage was observed within the lesion during the 1-month follow-up MRI. Dynamic contrast enhanced imaging showed mild increased perfusion of the peripheral area. Magnetic resonance spectroscopy showed an increased lactate/lipid peak of 1.3 ppm (**Figure 2**). Computed tomography (CT) of the sella was performed preoperatively (**Figure 3**), which revealed bone remodeling in the frontal and lamina papyracea adjacent to the lesion.

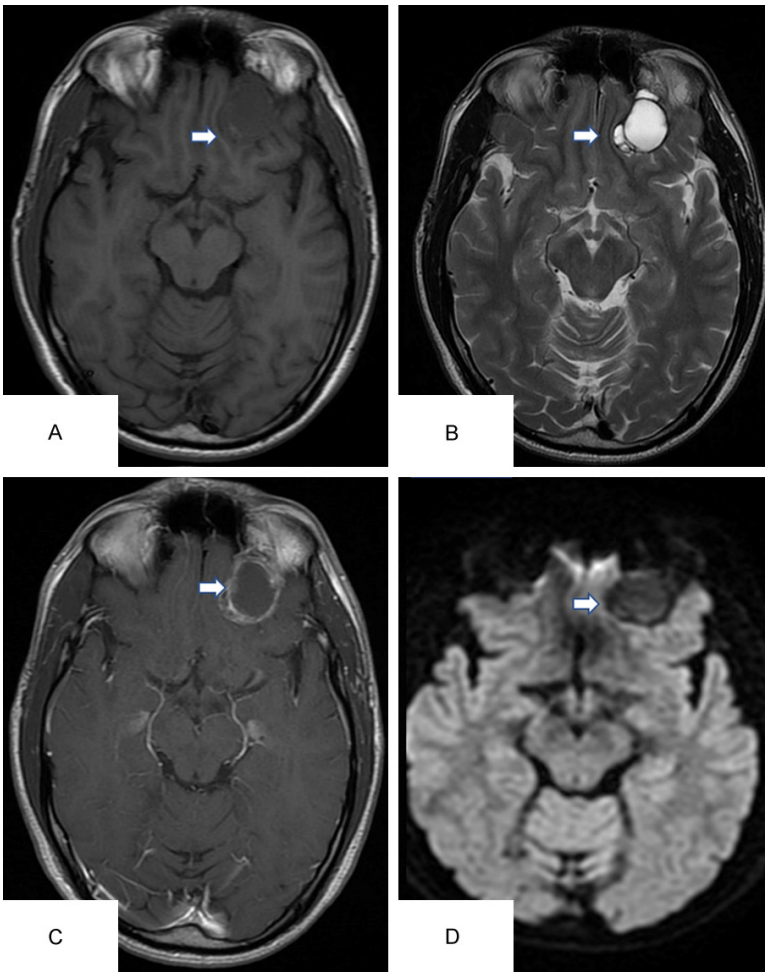

Figure 1. Initial magnetic resonance imaging of the brain of a 24-year-old man with psammomatoid ossifying fibroma. A. T1-weighted (T1W) image shows a hypointense lesion in the left frontal extra-axial space with faint T1 hyperintensity, suggesting a small hemorrhage (arrow). B. T2-weighted (T2W) image shows the lesion hyperintensity with a hypointense rim (arrow). Small lesions with the same characteristics are also visible. C. Contrast enhanced T1W image shows peripheral enhancement of the lesion (arrow). D. Diffusion weighted image shows no diffusion restriction (arrow).

Based on these imaging findings, the initial differential diagnoses were cavernous lymphangioma, hemangioma, venous varix, and aneurysmal bone cyst.

Combined open and endonasal surgery was performed, and our final diagnosis was a psammomatoid OF. After complete resection, the patient exhibited none of the previously reported symptoms. Microscopically, the tumor was a fibro-osseous neoplasm consisting of a hypercellular stroma and irregular-shaped bony trabeculae (**Figure 4A**). The tumor exhibited partial hemorrhage and cystic degeneration, resulting in the aggregation of numerous red blood cells and hemosiderin-laden macrophages (**Figure 4B**). Innumerable psammomatous bodies were identified in stellate and spindled stromal cells (**Figure 4C**). These tumor cells were negative for epithelial membrane antigen (EMA), smooth muscle actin (SMA), S100 and cluster of differentiation-34 (CD34), although positive for vimentin (**Figure 4D**). The Ki-67 proliferating index was approximately 5%. After the patient was discharged from the hospital, no recurrent lesions or symptoms were observed at the follow-up visit.

Discussion

OFs are categorized as three different types: COF, JTOF, and JPOF. JPOF is characterized by the presence of multiple uniform, small, round osteoid masses with concentric calcifications patterns, giving it a psammoma body-like appearance [3]. These three entities have different predilection sites. JPOF tends to occur in the sinonasal area, whereas COF and JTOF develops in the gnathic areas (COF favors the mandible; JTOF the maxilla) [4]. JPOF and JTOF have a male predilection, whereas COF has a female predilection.

A rich myxofibrous cellular stroma that promotes septal growth in the paranasal sinuses constitutes the pathogenesis. A total of 262 cases of JPOF have been previously identified in the literature [5]. Due to the rarity of JPOF, its risk factors remain unknown; however, a new tumor suppressor gene (*HRPT2*) mutation has been reported in OF . However, this genetic alteration is absent in JPOF, and is considered specific to COF [4].

Studies have shown that JPOFs appear in young people (mean age, 16-33 years) and display a slight male predilection. The nose and paranasal sinuses are the most common sites, with only 10% of cases occurring in the calvaria. The

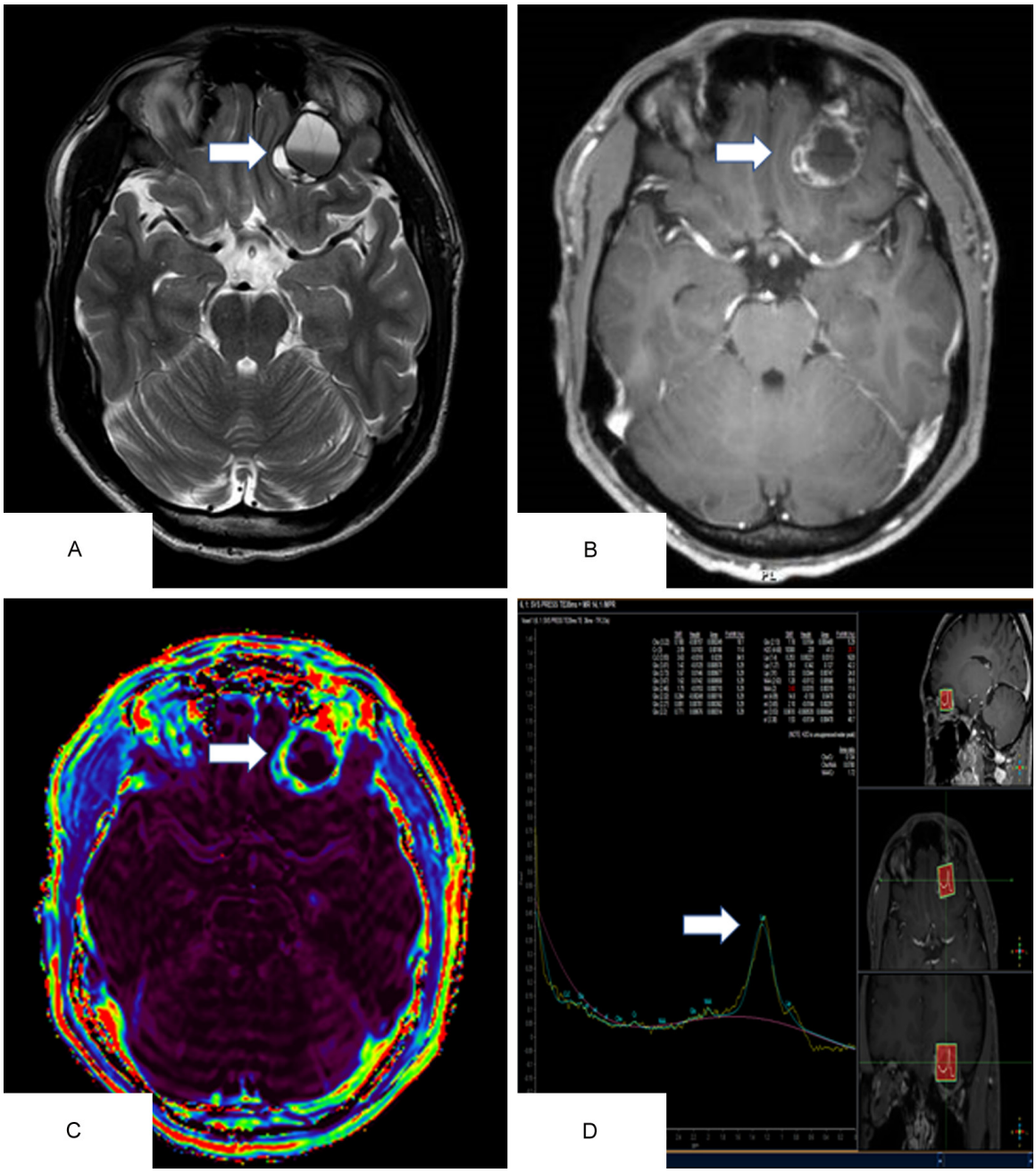

Figure 2. Follow-up brain magnetic resonance imaging of a 24-year-old man with psammomatoid ossifying fibroma. A. T1-weighted (T1W) image shows the cystic lesion compressing the left orbitofrontal lobe with newly visible multiple blood-fluid levels (arrow). B. Contrast-enhanced T1W image shows peripheral enhancement of the lesion (arrow). C. Dynamic contrast enhanced perfusion shows no increase in central perfusion with only mildly increased peripheral perfusion (arrow). D. Magnetic resonance spectroscopy shows an increased lipid/lactate peak (arrow).

most common clinical symptoms are proptosis and visual disturbance due to the location of the JPOF. Other symptoms include headache, facial swelling, and nasal stuffiness.

JPOFs may present as one of three imaging patterns on CT: (1) dense and homogeneous ; (2) ground glass mural; or (3) radiolucent center with a thick outer. The bony wall is isointense and hypointense on T1- and T2-weighted images, respectively. Homogeneous or heterogeneous enhancement is seen with contrast, although peripheral and septal enhancement can also be observed in the cystic areas.

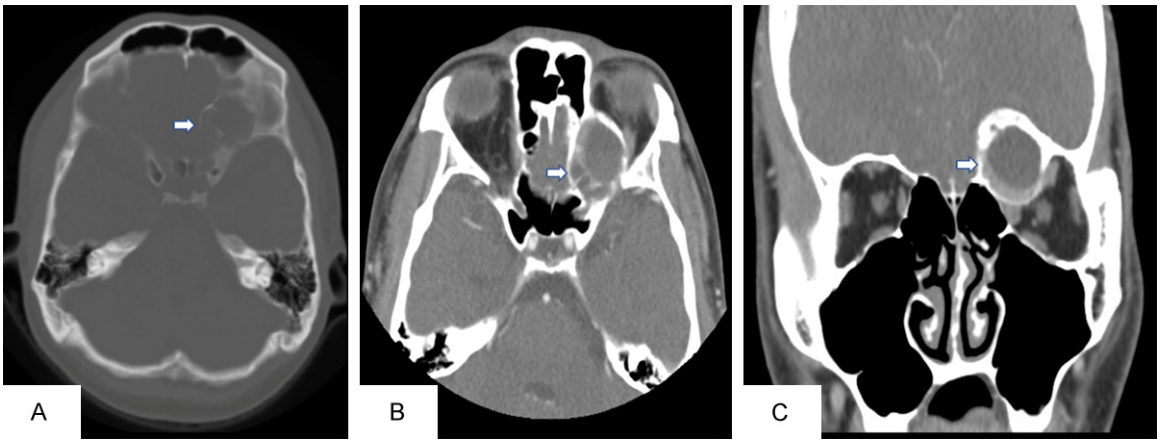

Figure 3. Preoperative brain computed tomography (CT) of a 24-year-old man with psammomatoid ossifying fibroma. A. Non-contrast CT shows an ovoid lesion with peripheral bony remodeling (arrow). B. Enhanced CT shows mild peripheral enhancement (arrow). C. Enhanced CT shows peripheral mild enhancement with left extraocular muscle compression and frontal bone remodeling (arrow).

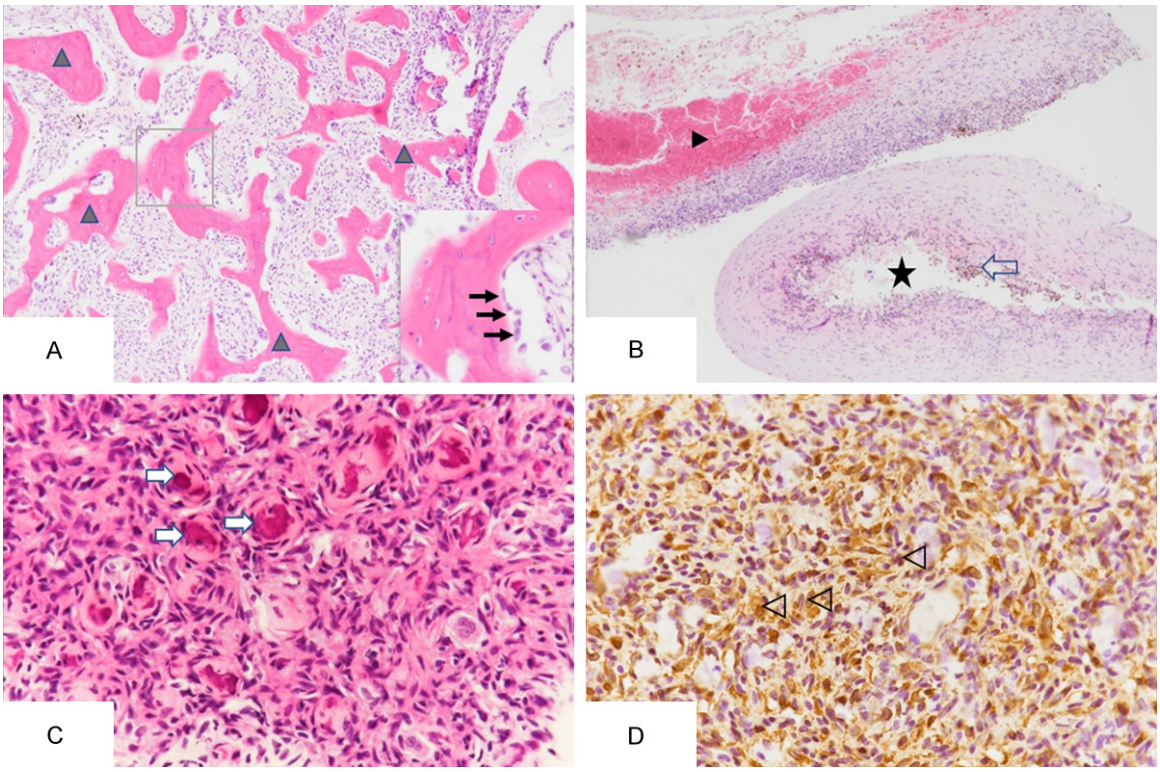

Figure 4. Hematoxylin & eosin (H&E) staining and immunochemistry of the lesion. A. Tumor forms curvilinear trabeculae (arrowheads) with osteoblastic rimming (black arrows) (H&E, ×100). B. Hemorrhage (black arrowhead) with hemosiderin pigment deposition (open arrow) and cystic degeneration (black star) are identifiable (H&E, ×100). C. The hypercellular stroma consists of spindled and stellate fibroblastic cells. Numerous calcifications can be seen in the center of eosinophilic cementum-like spherules (white arrows) that are embedded in the stroma (H&E, ×400). D. These tumor cells are positive for vimentin (open arrowheads) and negative for, smooth muscle actin, S100, epithelial membrane antigen, and cluster of differentiation-34. The Ki-67 proliferating index is approximately 5%.

The uniform fibroblastic spindle cells and extensive psammomatoid calcification may be cause the homogeneous signals on MRI and ground-glass opacity on CT. However, cyst formation

resulting from secondary aneurysmal bone cysts and hemorrhage reportedly occurs in up to 50% of patients. Our patient exhibited an increased lactate/lipid peak, likely owing to cystic changes. JPOF ossicles have an osteoid rim on pathological examination. The non-osseous component is either loosely or densely cellular, with proliferation of multinucleated, osteoclast-like, giant cells [3]. Although the immunohistochemical characteristics remain unclear, our finding of vimentin, SMA, CD10, and CD34 positivity with S-100 and EMA negativity concurs with those of the cases described in the literature [6].

JPOF is generally treated with total resection with a favorable outcome; however, the endonasal endoscopic approach is a potential alternative with equivocal safety. Radiotherapy is not recommended owing to the risk of malignant transformation. The recurrence rate is 30-56%, and recurrence can occur anywhere from 6 months to 19 years after treatment [6, 7]. When complete removal is not possible, close imaging follow-up without adjuvant therapy should be considered [1].

Imaging characteristics and tumor location are helpful in the differential diagnoses of fibrous dysplasia, JTOF, and osseous dysplasia [8]. Fibrous dysplasia shows ground glass opacity and can arise from any skeletal system, thereby excluding JTOF and OF. Fibrous dysplasia has no clear border, is self-limiting, and cannot be completely separated from the adjacent normal region, whereas JPOF tends to grow slowly. JTOFs appear in the maxilla or mandible and may have scattered calcification. Osseous dysplasia arises from the periapical regions of the jaw with varying composition. Psammomatoid meningioma can be difficult to distinguish from JPOF because it shows focal bone expansion and may have a similar ground glass appearance. Histologically, psammomatoid meningioma lacks osteoclasts and osteoblasts, and psammoma bodies are randomly distributed and not as homogeneous as JPOFs. Notably, JPOFs may include areas of aneurysmal bone cyst formation. Therefore, differentiation from aneurysmal bone cysts may be challenging.

In conclusion, despite its rarity, JPOF should be considered when multicystic lesions with peripheral enhancement near the orbit exhibit a progressive inner hemorrhage base. These imaging patterns along with demographics may guide a more accurate pre-operative diagnosis. The differential diagnoses include fibrous dysplasia, psammomatoid meningioma, and aneurysmal bone cyst.

Disclosure of conflict of interest

None.

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