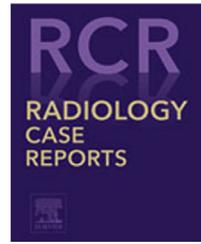


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## Case Report

# Juvenile psammomatoid ossifying fibroma: A radiolucent lesion to suspect preoperatively

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## ABSTRACT

We present the cases of 2 expansive juvenile psammomatoid ossifying fibromas from sinonasal origin. Our first patient presented with a fronto-ethmoidal mass invading the orbit and the cranial base and had a bicoronal approach for tumor removal. The second patient also had orbital involvement and underwent an endoscopic surgery. Complete resection of juvenile psammomatoid ossifying fibromas is paramount to avoid recurrence, thus preoperative recognition of their characteristic thick outer mantle and radiolucent core on imaging is key, but can be challenging. We herein discuss and propose a novel algorithm of differential diagnoses of facial bone lesions based on radiologic appearance.

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## Introduction

According to the newly released 2017 WHO classification, there are 3 variants of ossifying fibromas: cemento-ossifying fibroma, juvenile trabecular ossifying fibroma, and juvenile psammomatoid ossifying fibroma (JPOF) [1]. Previously con-

sidered a tumor of odontogenic origin (2005 WHO classification), JPOFs are now classified as a distinctive type of ossifying fibroma, under benign fibro- and chondro-osseous lesions [2]. Due to their distinctive histology and location to the paranasal sinuses, the orbit and the nasal cavity, they are distinguished from cemento-ossifying fibromas occurring in the jaw, arguably from periodontal ligament (thus odontogenic

*Abbreviations:* JPOF, juvenile psammomatoid ossifying fibromas; JTOF, juvenile trabecular ossifying fibroma.

**Competing Interests:** The authors have declared that no competing interests exist.

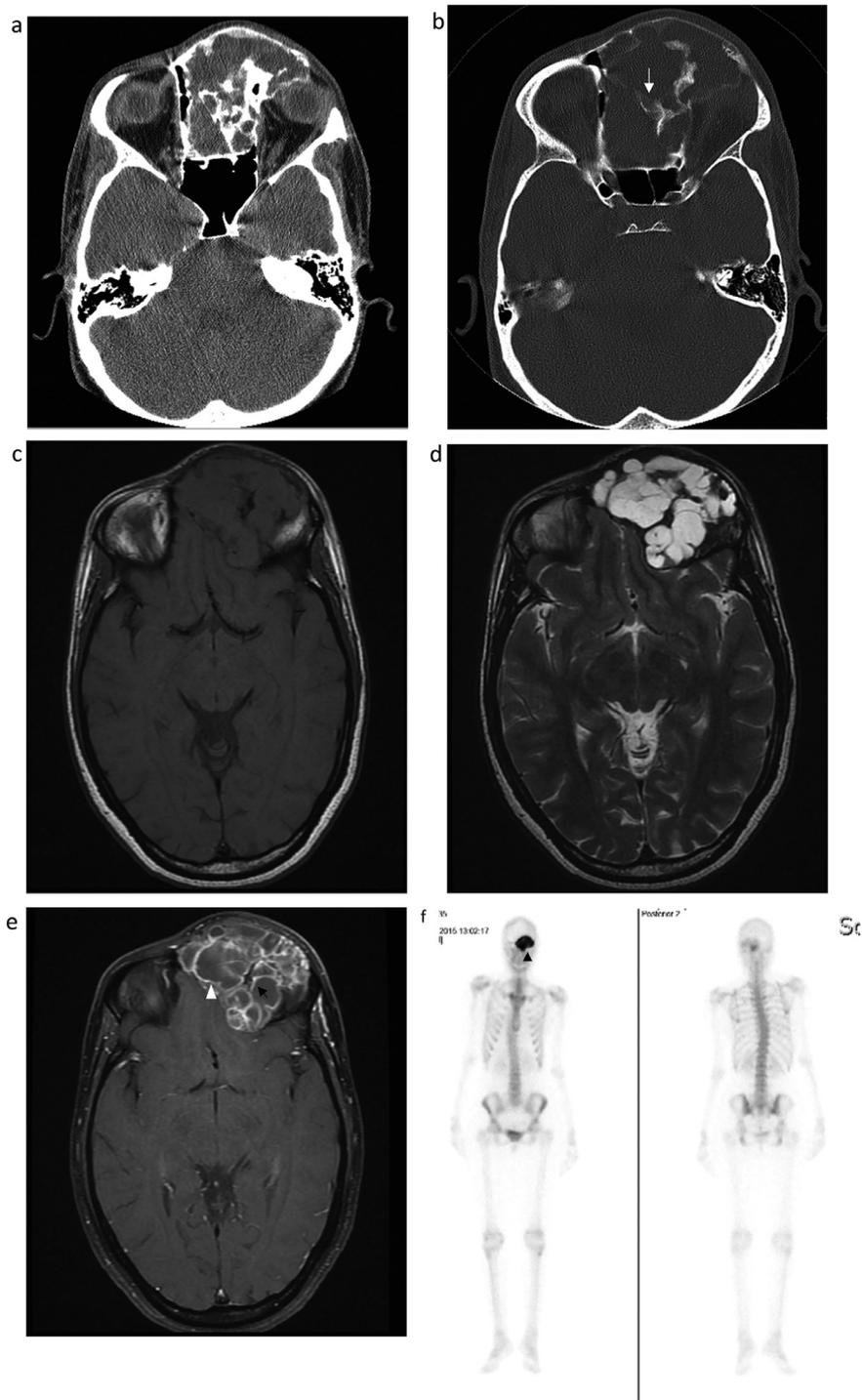
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**Fig. 1 – (a and b) Axial CT images without contrast displayed in a tissue window (a) and a bone window (b) depict a well-circumscribed low density (40 HU) frontal sinus mass with tissue component and calcified septation with ground-glass opacity (white arrow). (c) Axial T1-weighted image, (d) axial T2-weighted image and (e) postgadolinium injection T1-weighted image. MR images show a multiloculated expansile mass centered in the left frontal sinus. (c and d) Internal composition of the lesion is mainly in hyposignal in T1-weighted image and hypersignal in T2-weighted image. (e) MR images show avid septations (black arrow) and peripheral rim enhancement (white arrowhead). (f) Tardive phase of pancorporal osseous scintigraphy (2–4 hours postinjection of Technetium Tc-99 m medronate) demonstrates accumulation in a left frontal mass (black arrowhead).**

origin) [3]. JPOFs are rare benign tumors, but they can be locally aggressive and invade vital structures such as the orbit and the cranial base.

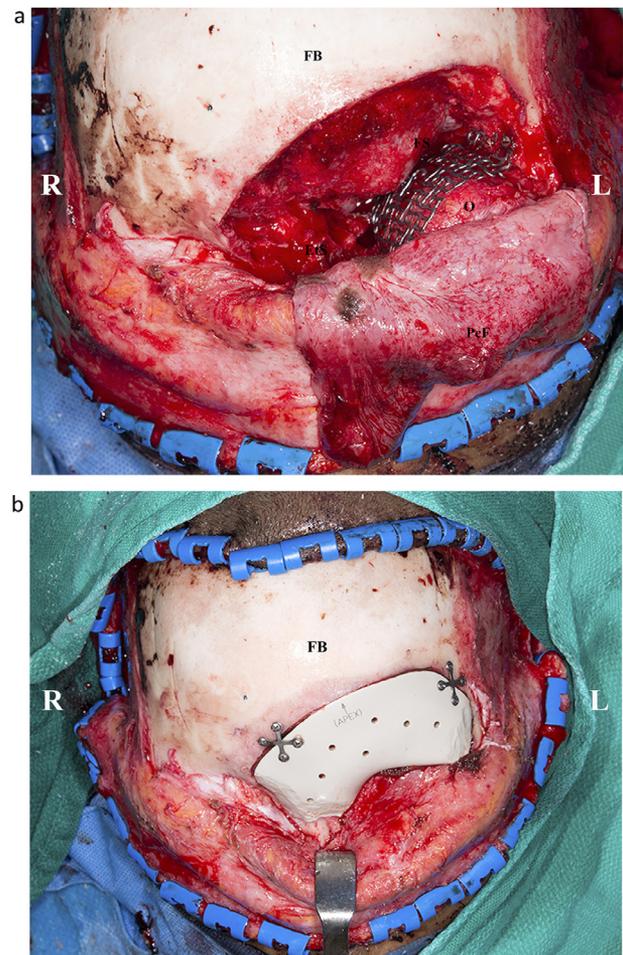
We herein present 2 cases of expansive JPOF located in the paranasal sinuses, with orbital involvement.

### Case report 1

A healthy nonsmoking 23-year-old female was referred to our otorhinolaryngology clinic by an outside Otorhinolaryngology for a recurrent fronto-ethmoidal mass diagnosed as a mucocele, and operated twice abroad. On medical history, she complained of a left frontal mass present during childhood that slowly progressed over years. She had intermittent headache. On physical examination, a bulging left frontal mass was noted, with inferolateral displacement of her left eye. She had no visual symptoms. Flexible endoscopy showed left nasal cavity synechia confirming her previous endoscopic sinus surgeries. On sinus computerized tomography (CT) scan, an expansive frontal sinus lesion (6.5 cm × 5.4 cm × 4.5 cm) with extension to ethmoidal cells was noted (Fig. 1). The lesion had calcifications. The superior orbital and the anterior and the posterior tables of the frontal sinus showed deformity, with suspicion of structural invasion. Magnetic resonance imaging (MRI) confirmed the previous findings and reported the presence of septations within the mass. Biopsy was performed and compatible with a JPOF. In November 2016, a multidisciplinary surgery was done (otorhinolaryngology, maxillofacial surgery, and neurosurgery). Peroperatively, frontal sinus bone, orbital roof, and cranial base were invaded, without intradural expansion. The patient underwent a near total excision of the benign tumor (bicoronal approach), reconstruction of the orbital roof with a 3D titanium grill and reconstruction of the anterior table of the left frontal sinus with a PEEK implant and a pericranial flap (Fig. 2). No cerebrospinal fluid leak was noted during the intervention. Her postoperative course was unremarkable (discharged on postoperative day 3). The patient is followed with CT scan every 6 months, and the residual tumoral tissue located at the ethmoid cells and the frontal bone is stable (Fig. 3). Her headaches have resolved.

### Case report 2

A 29-year-old male, with a medical history of pulmonary embolism, was referred to the otorhinolaryngology clinic by an outside otorhinolaryngologist for a right ethmoido-sphenoido-orbital mass incidentally found on head CT scan. His surroundings noted subtle exophthalmia. He complained of blurry vision. Sinus CT scan showed a 3.7 cm × 2.4 cm × 2.6 cm mass departing from the sphenoid sinus, with dehiscence of his sphenoidal planum, extending along posterior ethmoidal cells, penetrating the lamina papyracea without transgression of the periorbit, but creating a mass effect on his right orbital content. Ophthalmology evaluation showed optic nerve compression (Fig. 4). Biopsy of the lesion was compatible with JPOF. The patient underwent complete endoscopic re-

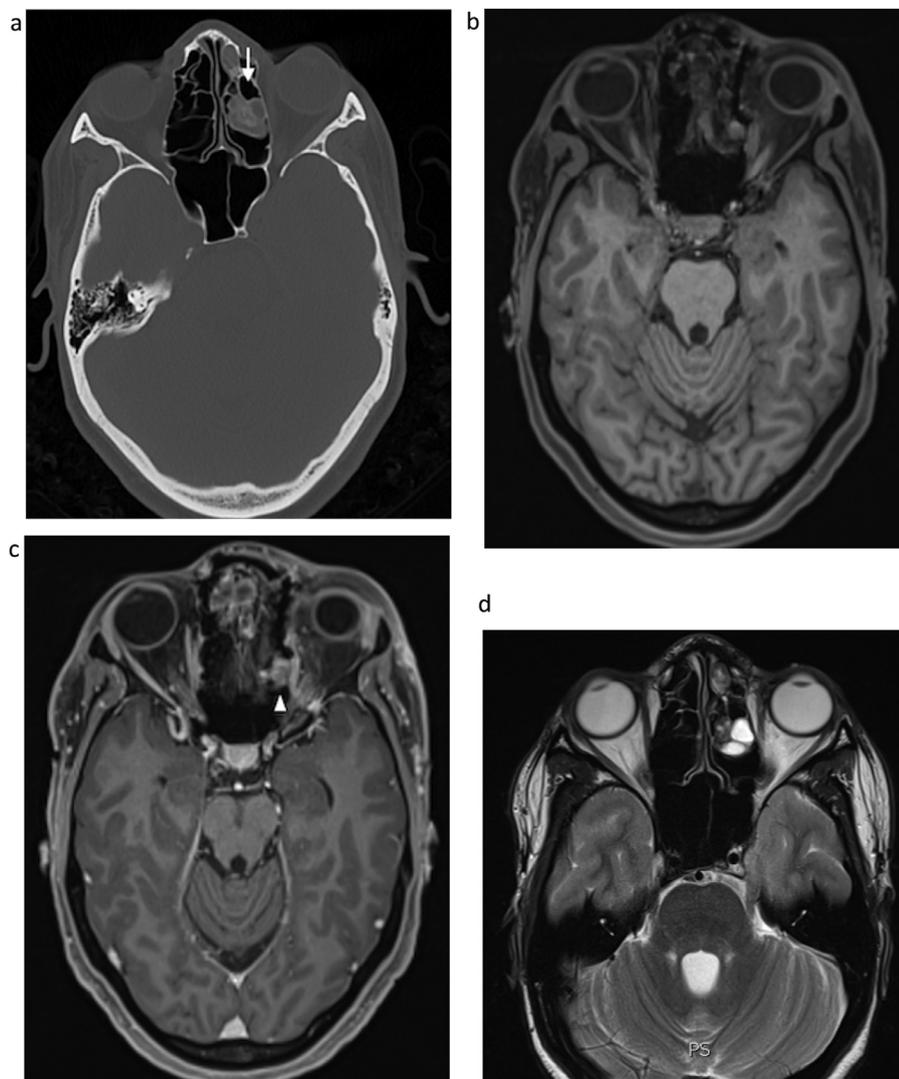


**Fig. 2 – (a) Reconstruction of the left orbital roof with a 3D titanium grill and (b) reconstruction of the anterior wall of the left frontal sinus with a PEEK implant. EtS, ethmoid sinuses; FB, frontal bone; FS, frontal sinus; L, left; O, orbital content; PcF, pericranial flap; R, right.**

section of the sphenoid tumor. Peroperatively, the tumor was found to invade and fuse with the laminae papyracea, which was also resected with the tumor. He was discharged on postoperative day 1. His postoperative course was unremarkable. His proptosis and orbital compression were relieved and postoperative visual acuity was normal. No complications or recurrence was noted on follow-up.

### Discussion

JPOFs mainly affect children and young adults (mean age 20 years old), with a male predilection [4,5]. On the other hand, conventional ossifying fibromas are more common in females and occur between the second and fourth decade (mean age 35 years old) [2]. Aggressive growth and higher recurrence rate in JPOF have been associated with younger age [4]. Johnston et al suggested that JPOFs are derived from



**Fig. 3 – One year post-op, (a) axial CT images without contrast displayed in a bone window depict a well-circumscribed ethmoid sinus mass with tissue component and residual ground glass opacity (white arrow). Two years post-op, (b) axial T1-weighted image, (c) axial T1-weighted with gadolinium image and (d) T2-weighted image depict sequelae of left frontal orbit cranioplasty with a stable residual lesion components around the crista galli, right frontal recess, left ethmoid sinus and left orbital roof with left extraconal orbital extension (white arrow head).**

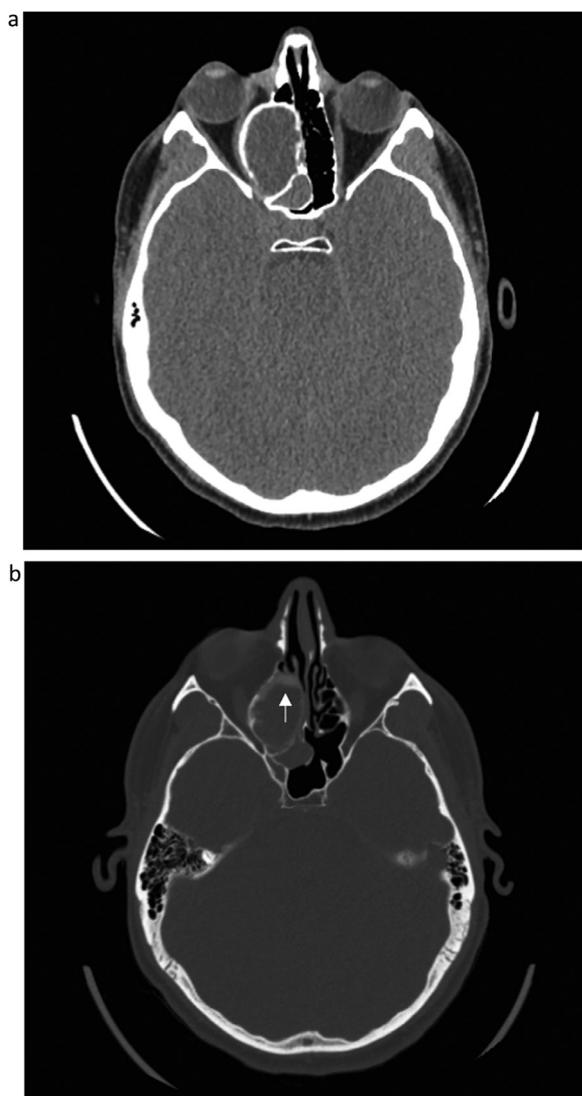
overproduction of myxofibrous cellular stroma involved in paranasal sinuses septa growth [6].

Paranasal sinuses are the sites of predilection for JPOFs. Other sites that can be affected include the orbit, the fronto-ethmoidal complex, the nasal cavity, the maxilla, the mandible, and the calvaria [4,5,7].

JPOF usually presents with localized unpainful facial swelling or craniofacial deformity. When symptomatic, symptoms often result from local tumoral extension. Visual complaints include proptosis, eyeball displacement, lagophthalmos, restricted eye movements, and decreased visual acuity. Sinusal invasion can manifest as recurrent sinusitis or nasal obstruction [4,5]. Headaches have also been reported [8]. The clinical course is variable. Typically, symptoms progress over weeks to months, but lesions progressing over decades have also been reported [9].

Radiologic imaging, CT scan or MRI, is necessary to determine the extension of the tumor and the bony involvement. JPOF usually presents as a radiopaque uni- or multiloculated lesion with well-defined corticated borders, without soft tissue involvement. The inner core can be radiolucent, radiopaque, or mixed. CT scan may suggest cystic lesions like mucocèles as primarily evoked in the presented cases [4].

We usually think about fibro-osseous lesion as having bone or soft tissue signals, not radiolucent lesions. According to Owosho et al, there are 3 different radiological patterns of JPOFs on CT scans: (1) an outer thick mantle with a radiolucent core; (2) a single ground glass mural nodule; or (3) a solid, homogenous radiopacity [10]. Psammomatoid ossicles within the lesion can also present as radiotransparent ground glass opacities [4].



**Fig. 4 – (a and b) Axial CT without contrast image in a tissue window (b) and a bone window, show a low density (30 HU) lentiform expansile lesion within right ethmoidal sinus, with an internal septa, well-defined borders with ground-glass opacity (white arrow).**

On MRI, the bony wall of the lesion (calcification rim) is isointense on T1-weighted images and hypointense on T2-weighted images. MRI can also demonstrate bony shell enhancement with contrast, suggesting tumoral tissue rather than reactive hyperostosis [11]. Solid components are usually isointense compared to adjacent muscles on T1-weighted images and iso- to hypointense on T2-weighted images [12]. After administration of intravenous gadolinium contrast, solid components might show heterogeneous enhancement, while cystic areas may present with peripheral and septal enhancement [12].

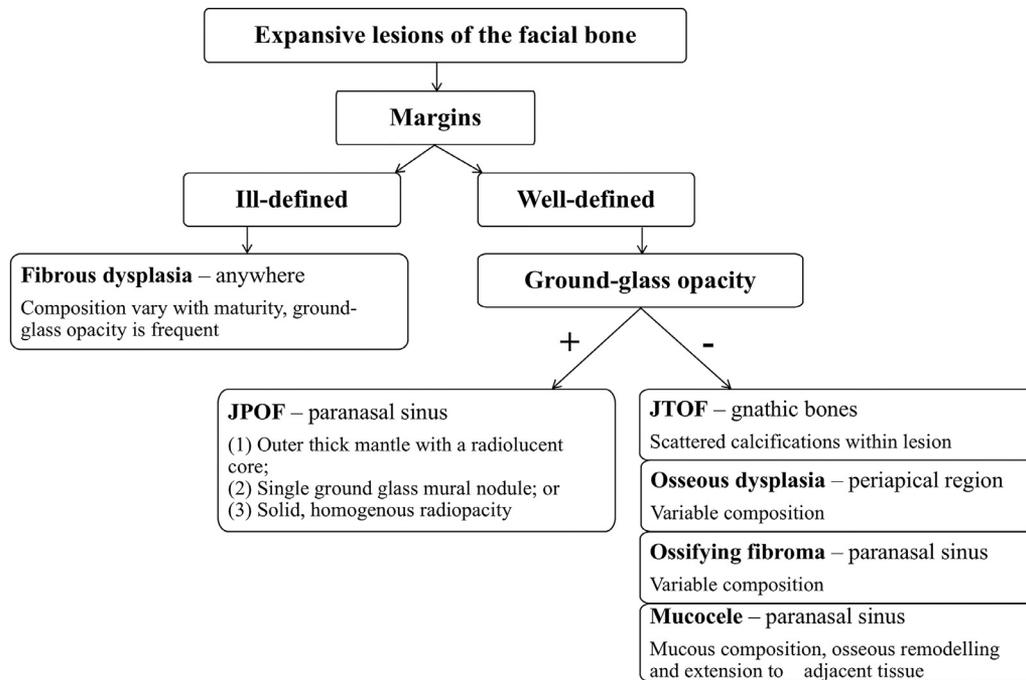
Microscopic findings are characterized by fibroblastic stroma, loose, or intensely cellular with minimal collagen, containing spherical collections of calcium. These small ossicles found in the fibroblastic stroma resemble psammomas'

round bodies, but unlike them, they can show loosely distributed cells and are curved without being perfectly spherical. These ossicles may fuse to form trabeculae, or may be grouped by their thick irregular collagenous rim. Aneurysmal bone cyst areas can result from trabeculae of woven bone as well as lamellar bone, pseudocystic stromal degeneration, and hemorrhages. These aneurysmal bone cyst areas result in multiple cysts and fluids level, which give JPOFs their typical radiological imaging [2,10].

On immunohistochemistry, JPOFs show positivity for vimentin, SMA and CD10. They lack the expression of CD34, S-100 proteins, and cytokeratins. Staining for epithelial membrane antigen (EMA) can sometimes be positive in JPOF and should be used carefully to distinguish the JPOF (usually negative for EMA) from extracranial psammomatous meningioma (positive for EMA) [2,13].

Diagnosis is based on histopathology and imaging. The differential diagnosis of JPOF includes all expansive lesions arising from the facial bones, such as fibrous dysplasia, juvenile trabecular ossifying fibroma (JTOF), mucocele, osseous dysplasia, or other types of ossifying fibroma. Narrowing the differential diagnostic can be done according to the tumoral margins characteristics, the presence of ground-glass opacity and the location of the tumor (Fig. 5) [4,10,12]. Fibrous dysplasia has ill-defined margins, which is a distinguishing feature from other expansive lesions. Fibrous dysplasia can be present anywhere on the skeleton and have ground glass opacities depending on the maturity of the lesion. The presence of ground-glass opacity allows exclusion of JTOF, ossifying fibroma and mucocele, which all lack this composition. Mucocele may become large lesions with smooth remodeling walls and bony changes, with extension in adjacent tissue. Furthermore, the location of the lesion also helps with the diagnosis. JTOF typically arises from the maxilla or mandible (gnathic bones), osseous dysplasia occurs in periapical regions of the jaw, and mucocele and JPOF, in sinonasal regions [5]. JTOF may have scattered calcifications and also occur in younger patients. Osseous dysplasia and ossifying fibroma have variable composition. Psammomatous calcifications of JPOF are usually absent in the trabecular variant of juvenile ossifying fibroma [10]. Other rare malignant bone tumors of the childhood must be eliminated, such as osteosarcoma, chondrosarcoma and Ewing's sarcoma [4].

Surgical care often commands multidisciplinary management, including otolaryngology, neurosurgery, ophthalmology, and maxillofacial surgery, due to tumoral extension and invasion of adjacent structures [14]. Solely endoscopic approaches are possible in selected patients, depending on the tumoral extension, but open surgeries might be necessary for total tumoral resection and optimal local control [15]. Total tumoral resection must include resection of the thick outer mantle of the tumor. If JPOF is not suspected preoperatively, one might inadvertently proceed to an incomplete tumoral excision. Postoperative recurrence is common, especially with partial tumoral excision. One should not compromise complete tumoral excision to avoid open surgery, as complete removal is superior to curettage, and incomplete or partial excision [2,4,8]. Incomplete resection is reasonable when the tumor is closed to vital structures, like in our first case. No malignant transformation or metastatic disease was reported



**Fig. 5 – Algorithm of differential diagnoses of expansive lesions of the facial bone according to radiologic appearance.**

[2]. A follow-up is needed to monitor recurrences or tumor progression, especially with incomplete resection. Postoperative radiation therapy has no role in the current therapeutic algorithm.

## Conclusion

We present the cases of 2 expansive JPOF with orbital involvement and a literature review. JPOFs are now considered a distinct entity from convention ossifying fibromas, due to their histology and location to the paranasal sinuses. On radiologic imaging, JPOFs have a characteristic thick outer mantle with a radiolucent core appearance, which makes their diagnosis challenging. Although benign, they can be locally aggressive. It is important to suspect the diagnosis of JPOF preoperatively, as they should be treated surgically, with emphasis on complete resection for better local control and with a multidisciplinary approach.

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