



Juvenile psammomatoid ossifying fibroma of the ethmoid sinus successfully treated by endoscopic sinus surgery: a case report

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Background: Juvenile ossifying fibromas (JOF) are benign osteofibrous tumors very rarely located in paranasal sinus. The diagnostic and therapeutic tools applied in sinonasal inflammatory pathologies and other tumors fail to address a specific management in this rare pathology. Currently, the only available treatment is surgery. Endoscopic sinus surgery (ESS) allows a complete but minimally invasive approach compared to the open technique, especially for the minor demolition on growing bone structures.

Case Description: We describe a case of a 13-year-old boy presenting in the Emergency Department for a head trauma. The computerized tomography (CT) revealed an ethmoid-maxillary bony mass suspected to be a juvenile ossifying fibroma. The patient underwent ESS. Histological examination confirmed a juvenile psammomatoid ossifying fibroma (JPOF). A revision surgery with the same technique was required after 7 months due to relapse of the tumor. He did not develop postoperative complications. The 3-years follow-up did not show any signs of relapse.

Conclusions: Paranasal JOF is a challenge for the ear-nose-throat (ENT) surgeon. The principles of local disease control for relapse reduction are different from inflammatory diseases and other paranasal tumors. Severe organ damage and recurrences are possible if caution is not observed. Specific guidelines about management are required on the basis of the shared experiences in the few cases reported in literature.

Keywords: Juvenile ossifying fibroma (JOF); endoscopic sinus surgery (ESS); paranasal sinus; case report

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Introduction

Ossifying fibroma is a benign tumour first described by Menzel in 1872, characterized by the replacement of normal bone tissue with a fibrous cellular stroma that contains numerous foci of mineralization (1). It generally occurs in patients in their second to the fourth decade, women are more commonly affected than men and there is no evidence of inheritance (2). Ossifying fibromas of the head and neck

are rare, mostly affecting the mandible/maxilla and rarely the orbit, the skull base, and the paranasal sinuses (3). The term "juvenile" is used when patients under 15 years old are affected.

The juvenile variant tends to be locally aggressive, with high risk of recurrence if incompletely removed. Juvenile ossifying fibromas (JOF) are usually divided into two subtypes: trabecular (JTOF) and psammomatoid (JPOF).

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Histologically, JTOF is characterized by the deposition of minerals leading to the formation of trabeculae of woven bone, while JPOF shows fibrous stroma and spheroidal calcifications called “psammoma-like bodies”. The mean age of the trabecular type is usually younger than the psammomatoid (4). A recent systematic review reported a total 143 cases of paranasal JOF, 12 were trabecular and 131 psammomatoid (5), so paranasal JOFs are psammomatoid in most cases. Clinical management do not differ between the two types. Paranasal JPOFs occur more frequently in frontal and ethmoid sinuses, from which it can extent to the orbit and the anterior cranial fossa (4,5). The clinical manifestations are due to the bony expansion, and may include nasal obstruction, facial swelling, proptosis, and headaches. Nonetheless, JPOF are asymptomatic in 96.5% cases (4). When properly treated, the prognosis of paranasal JPOF is good, however a recurrence rate 27.8–56% is reported in literature mainly due to the infiltrative nature of the tumour borders (4,5). The treatment is surgical excision by open, endoscopic, or combined techniques, depending on the location and extension of the pathology (5,6). We report a case of a 13-year-old boy with an ethmoid-maxillary JOF accidentally diagnosed by cranial computerized tomography (CT) and successfully treated by sinus endoscopic surgery. This case report was prepared following the CARE reporting checklist (available at <https://acr.amegroups.com/article/view/10.21037/acr-22-90/rc>) (7).

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki

Declaration (as revised in 2013). Written informed consent for publication of this case report and accompanying images was not obtained from the patient or the relatives after all possible attempts were made to contact them, and the article has been sufficiently anonymized to cause no harm to the patient or his family. We present the case of a 13-year-old Caucasian male transferred to the emergency room of Bambino Gesù Children’s Hospital from another hospital due to accidental facial trauma, after which maxillofacial CT was performed. The clinical history revealed that the patient had been suffering from a nasal obstruction for several months. He was in good health before the onset of the obstruction, with unremarkable medical, surgical, and family histories. The CT showed an expansive bony mass originating from the right ethmoid that expanded into the ipsilateral nasal cavity, the orbit, and the maxillary sinus (*Figure 1*). He was hospitalized and a magnetic resonance imaging (MRI) was performed to define the mass more fully and to clarify the involvement of the surrounding soft tissues, particularly the orbit. MRI excluded any intracranial and orbital complications.

Among the possible differential diagnoses, a giant osteoma or ossifying fibroma appeared to be the most compatible; histological typing was therefore considered necessary. The neoformation was biopsied under general anaesthesia with endoscopic control by 0° optics; the histological examination definitively confirmed a juvenile ossifying fibroma of the psammomatoid subtype (*Figure 2*). At the periphery of the lesion, the ossicles were fused and formed bone trabeculae showing numerous reversal lines (*Figure 2B*). Spherical small ossicles called psammoma-like bodies were founded in the sample examined (*Figure 2A*). There was focal pseudocystic haemorrhagic degeneration bordered by multinucleated giant cells similar to an aneurysmal bone cyst (*Figure 2C*).

A few days later, the patient underwent endoscopic surgery to remove the lesion. After infiltrating the lining mucosa, the mass was evacuated; the inside of the lesion appeared fibrous. The middle turbinate was removed, and the cleavage plane was collected from the nasal septum, after which the capsule delimiting the neoformation was excised, but, as a precaution, left in place in its lateral component in direct continuity with the papyrus lamina, which manifested signs of wear. No post-operative complications occurred. The patients did not complain any discomfort after the procedure.

One month after surgery, the first control CT scan showed residue of the “shell” of the lesion at the level of the

Highlight box

Key findings

- JPOF may be treated successfully with endoscopic sinus surgery (ESS).

What is known and what is new?

- Recurrence rate and anatomical preservation are the main challenges in the treatment of JPOF.
- ESS may guarantee preservation of structures even in revision surgery after JPOF’s recurrence.

What is the implication, and what should change now?

- Further research should investigate the real time CT-assisted ESS and the adjuvant interferon alpha therapy in the treatment of JPOF.

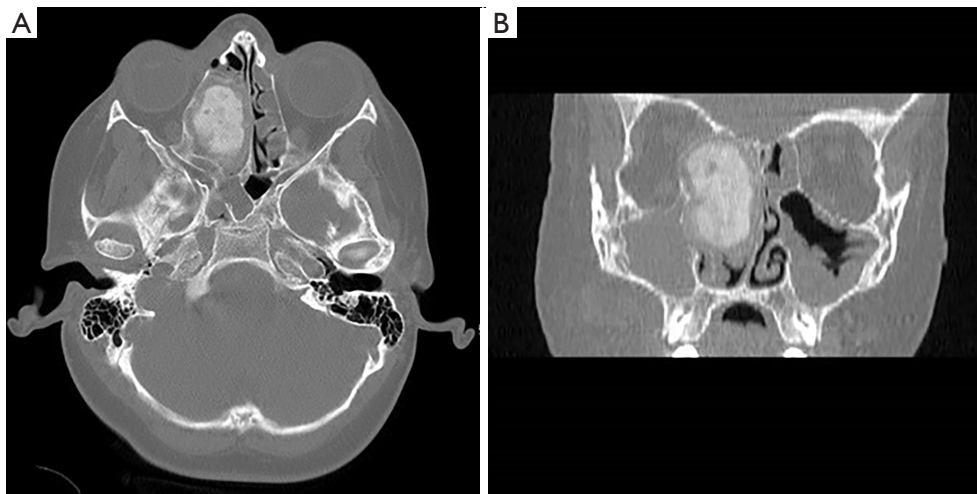


Figure 1 Preoperative maxillofacial CT showed an expansive bony mass of the right ethmoid expanding ipsilaterally into the nasal cavity, the orbit, and the maxillary sinus, and displacing the nasal septum. (A) Axial CT scan; (B) coronal CT scan. CT, computed tomography.

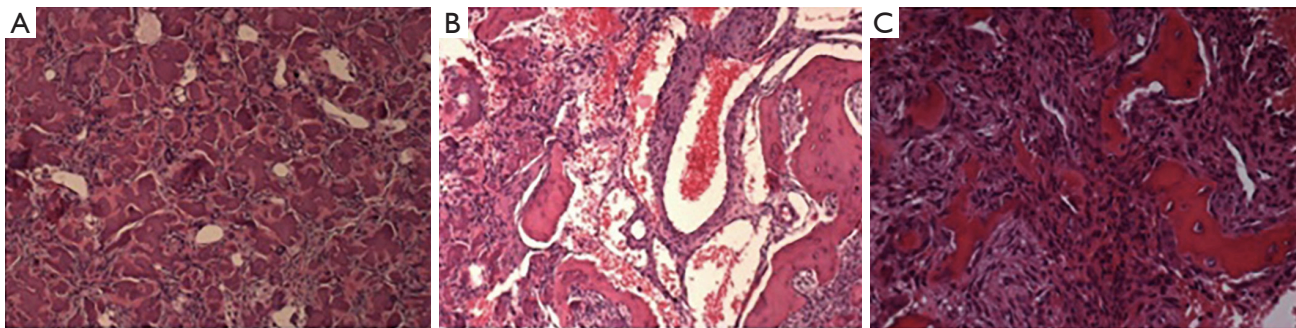


Figure 2 Histological HE pictures of the tumor. (A) Microscopic findings showed uniform spherical curved small ossicles—psammoma-like bodies, within a fibroblastic stroma of uniform stellate, or spindle shaped cells (HE, $\times 100$); (B) the ossicles are fused to form bone trabeculae, and reversal lines (HE, $\times 200$); (C) pseudocystic, haemorrhagic degeneration surrounded by multinucleated giant cells, similar to aneurismal bone cyst (HE, $\times 400$). HE, hematoxylin and eosin.

right orbital region. The patient's clinical conditions were stable and nasal endoscopic controls were normal.

The patient was stable up to the second postoperative month, until he experienced a headache and underwent an emergency CT which revealed an increase in the volume of the residual "shell" with convexity on the medial profile of the right orbit (*Figure 3*).

The patient was therefore hospitalized and underwent revision surgery with complete removal of the bone wall (newly formed "shell") laterally towards the orbit until Tenon's capsule was discovered, and up to the roof of the ethmoid. There were no postoperative complications, with

regular recovery and discharge.

Subsequent monthly endoscopic checks together with control CT (10 months after the last surgery) have highlighted the results of previous surgeries, without detecting any further residual disease, but only a minimal plus image of the profile of the medial wall of the orbit was likely reactive in nature (*Figure 4*).

Thirty-six months after the last surgery, the patient was asymptomatic with negative endoscopic controls for disease recurrence and was clinically stable. The patient is still continuing a regular follow-up, and no recurrence has been observed.

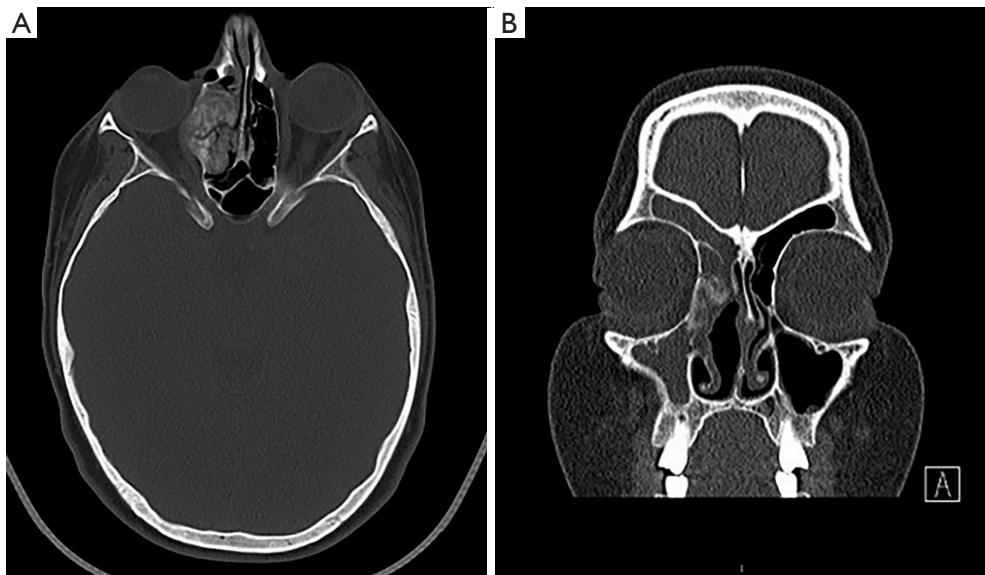


Figure 3 CT performed urgently in the emergency room. The axial (A) and the coronal (B) plans show an ethmoidal bony “shell” with convexity expanding on the median wall of the orbit. CT, computed tomography.

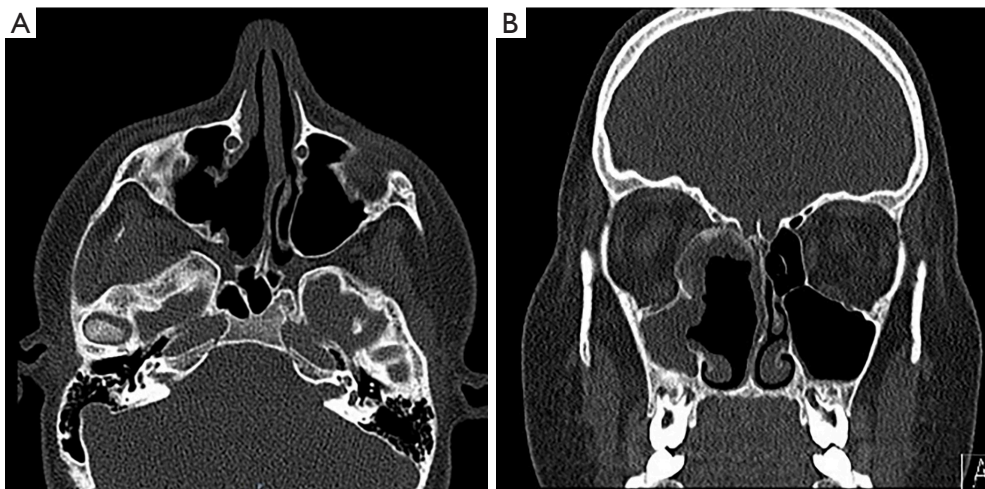


Figure 4 Follow-up CT images after ESS revision. The orbit’s median wall was preserved, ethmoid and maxillary sinuses were free from disease, with intact maxillary antrostomy. (A) Axial CT scan; (B) coronal CT scan. CT, computed tomography; ESS, endoscopic sinus surgery.

Discussion

JOF of paranasal sinuses is a rare pathology. Endoscopic treatment is accepted as applicable by now, however practical expertise may be not sufficient to guarantee radicality after the first surgery.

In this clinical case, we obtained the patient’s recovery and respected the integrity of the anatomical structures, although a surgical review was necessary.

A literature review revealed that paranasal osteofibrous lesions are often asymptomatic and incidentally diagnosed during investigations for other diseases, such as the head trauma in this case.

Only when the lesion protrudes into the nasal cavity it is possible to identify it endoscopically as a rounded swelling covered with an apparently healthy mucosa (5).

CT is the preferred imaging modality for this type of tumour and allows to distinguish the different subtypes of

osteofibrous lesions. The ossifying fibroma often appears as a mass with a well-defined edge, unlike fibrous dysplasia (FD), a disturbance of bone metabolism resulting in bone replacement with fibrous connective tissue. Radiologically the ossifying fibroma does not resemble a ground-glass opacity but has a mixed density between compact bone and fibrous tissue (8). In lesions of this type, MRI may help to evaluate the surrounding soft tissue involved, but a preoperative biopsy is always recommended in order to differentiate between the malignant forms (9,10). In addition to FD, other several pathologies should be taken into account in differential diagnosis: osteoblastoma, osteosarcoma, primary aneurysmal bone cyst, intraosseous cavernous hemangioma and eosinophilic granuloma. In doubtful cases, histological diagnosis may be performed with immunohistochemistry (5).

Open surgery (craniofacial resection) has been the gold standard of treatment of ossifying fibromas so far (11). This approach may still be valid for removing frontal sinus osteomas, which are the most frequent (about 57% of paranasal localization) and the most difficult to completely eradicate transnasally (12,13). Endoscopic techniques offer an optimal approach in all the other localization (ethmoidal, sphenoidal, and maxillary sinus) and also in frontal sinus when reachable through the ostium (14,15).

Recent studies have shown that with the advancement of endoscopic techniques, in expert hands, complete excision is possible by ESS in the majority of cases (13-16). Direct visualization of the anatomical structures avoids damage to surrounding structures. A real time CT navigation equipment can be used to guide surgeons in much safe and effective endoscopic procedures (12). The ESS allows a decidedly less invasive approach, which may be especially preferred in pediatric patients with still immature bony structures (13-16). In case of intracranial or orbital involvement, an external or combined approach is necessary for better control of the operative field, avoiding complications and reducing the risk of recurrence (17,18). The JOFs have an infiltrative behavior and incomplete excision on the tumour border causes relapses. Some Author suggests radical, *en bloc* resection in case of aggressive, rapid growing JOF, but a very extensive mass would require major demolitive surgery (18-20). This aspect in the case of benign tumor in childhood should be carefully evaluated. There are no effective medical therapies, because it is still unclear if the etiology is osteogenic (due to multipotential mesenchymal cells of the periodontal ligament), traumatic, or infective (20). Nonetheless, some author experienced a

protocol for aggressive JOF, consisting in surgical removal in combination with adjuvant interferon-alpha therapy (20,21). In both open and endoscopic surgery, enucleation should be followed by the curettage or osteotomy of the apparently healthy bone tissue near the margins of the tumour.

Conclusions

In the present case, a non-radical excision of the JOF led to a short-term relapse, confirming that complete excision of this type of lesion is essential for resolution. The endoscopic approach, therefore, should be the technique of choice, given the excellent access to the paranasal sinuses, greater preservation of the anatomical structures, the best postoperative course, and best functional and aesthetic results for a patient of developmental age. Further studies will provide more data about interferon therapy, in combination with endoscopic surgery, to avoid recurrences and demolition surgery, and to obtain radicality while conserving anatomical structures and function in young patients.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at <https://acr.amegroups.com/article/view/10.21037/acr-22-90/rc>

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Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised

in 2013). Written informed consent for publication of this case report and accompanying images was not obtained from the patient or the relatives after all possible attempts were made to contact them, and the article has been sufficiently anonymized to cause no harm to the patient or his family.

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