

Case Report

Juvenile ossifying fibroma of the orbit: a rare location

Pedro Clarós^{1*}, Sofia Polainas^{1,2}, Marta Fortuny³, Andres Clarós¹

¹Department of Otorhinolaryngology, Claros Clinic, Barcelona, Spain

²Department of Otorhinolaryngology, Hospital Garcia de Orta, Almada, Portugal

³Department of Oculoplasty, Claros Clinic, Barcelona, Spain

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*Correspondence:

Dr. Pedro Clarós,

E-mail: clinica@clinaclaros.com

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ABSTRACT

Osseous tumors in the craniofacial skeleton of young patients are not very frequent, and tumors involving the walls of the orbital cavity are even more infrequent. Despite being usually slow-growing, even small neoforations can have a local aggressive behavior, displacing and compressing vital structures, and so it is important to perform an early diagnosis in order to avoid the tumor to pose problems on the optic nerve and threaten vision. The case that we are presenting shows an ossifying fibroma in a 19-year-old male from Ethiopia, a benign lesion whose progressive growth caused proptosis and downward displacement of the left eye. Following an adequate radiological diagnosis, we were able to delimit the fibroma existence and location. The surgery was conducted in a conservative form, allowing the patient not to suffer from post-surgery malformations. Histopathological diagnosis was compatible with juvenile ossifying fibroma, psammomatoid variant, arising from an uncommon location – the roof of the orbit.

Keywords: Ossifying fibroma, Orbital tumor, Benign tumor

INTRODUCTION

Orbital tumors in young patients have a low incidence.¹ The most important problem is to know its stage of malignancy and its possible therapeutic problems. The radiological study and the pathologist's result will define the action to carry out as well as its prognosis.

Orbit surgery is a multidisciplinary approach in which otorhinolaryngologists, maxillofacial, neurosurgery, oculoplastic and head and neck specialists can compete. Undoubtedly the role of the pathologist in the detailed diagnosis is a key part. Histological diagnosis is not always easy and especially in these cases of psammomatoid variant tumors.

CASE REPORT

We report a case of a 19-year-old male from Ethiopia presenting with progressive proptosis and slight

downward displacement of the left eye for approximately one year. He had no associated pain or loss of visual acuity, no diplopia. On examination, no lesions were palpable on the edges of the left orbit.

Imaging studies (Figure 1) showed a lesion on the roof of the left orbit, originating from the frontal bone, with well-defined borders, 31×30×25mm of diameter, and multiple cystic-like small lesions inside. It caused downward displacement of the superior rectus muscle, and also of the optic nerve, that maintained its normal thickness and signal. Superiorly, the lesion produced compression of the left frontal lobe.

The patient underwent surgery for removal of the tumor. A bilateral supraciliary skin incision was made to the depth of the periosteum, and a superiorly based flat was secured with traction sutures to aid exposure (Figure 2). Downward detachment of the periorbita was followed by drilling of the upper left orbital bone rim that allowed exposure of the orbital side of the tumor, which extended

to the left frontal sinus. It was completely removed, maintaining the integrity of the upper cortical and the meninges. The orbital roof was reconstructed with a titanium mesh held in place by screws.

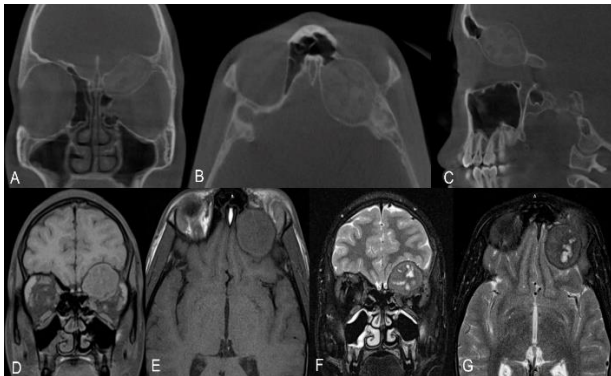


Figure 1: Preoperative CT scan, (A) coronal (B) axial (C) sagittal views, (D, E) MRI scan T1-weighted (F, G) T2-weighted showing a well-defined lesion on the left orbital roof.

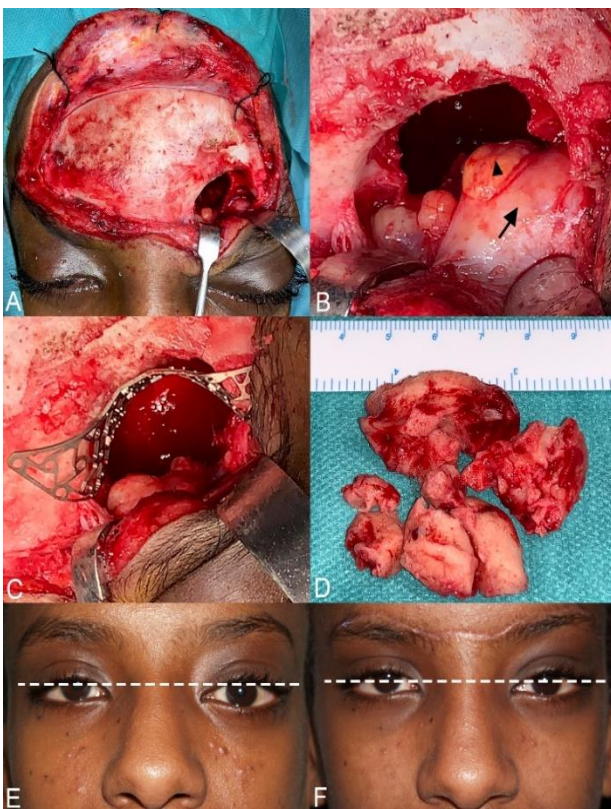


Figure 2: Images of the surgical intervention and the patient: (A) superiorly based flap, (B) tumor already removed, periorbita (arrow), orbital fat (arrowhead), (C) titanium mesh, (D) specimen of the tumor in pieces, (E) preoperative slight downward displacement of the left eye, (F) postoperative at 2 months, ocular globes aligned.

The histopathological exam showed a fibro-osseous tumor formed by a proliferation of elongated cells on a

collagen-rich stroma. Amongst these cells, a large number of psammomatoid structures of varying sizes, markedly mineralized, were observed. No cell atypia, necrosis or atypical figures of mitosis were observed. This is compatible with the diagnosis of juvenile ossifying fibroma, psammomatoid variant.

Two months postoperatively the patient had no symptoms, the CT scan showed no recurrence, and the titanium plate was in place (Figure 2).

DISCUSSION

Ossifying fibromas (OF) are uncommon fibro-osseous neoplasms that occur in the craniofacial skeleton, that, although benign, can have a local aggressive behavior.¹ There are two forms of the disease: a conventional form, also called cemento-ossifying, with an odontogenic origin that predominantly affects the mandibles of women in their third decade of life; and a juvenile form, rarer, that usually presents in men at a younger age, even during childhood. Juvenile Ossifying Fibroma (JOF) tends to grow more rapidly, and mostly affects the maxilla.^{2,3}

JOF manifests as a space-occupying lesion with a demarcation from the surrounding bone, as a painless swelling with rapid growth, that can cause facial asymmetry.^{1,4}

JOF has two different histopathological variants, the psammomatoid and the trabecular. The first is characterized by numerous small, rounded calcifications, called psammoma bodies, or sometimes “ossicules”, embedded in a cellular fibrous stroma; the second shows loose fibroblastic tissue with areas of collagen condensation, with subsequent deposition of minerals leading to the formation of trabeculae of woven bone.^{3,5} Psammomatoid JOF seems to be more prevalent, arises around paranasal sinuses and orbits, as well as the maxilla, is more aggressive and has a higher recurrence rate, whereas trabecular JOF mainly affects the maxilla and mandible.^{2,3,5}

Although the incidence of JOF is not known, a recent systematic review of JOF of the jaws and paranasal sinuses found 491 cases reported in the literature until march 2019.⁵

Orbital OF are very rare. Orbital involvement mostly occurs secondary to lesions that arise from the bony walls of paranasal sinuses or in the orbital plate of the frontal bone.⁶ Over 90 cases of OF involving the orbit have been described in the literature, and the vast majority of these cases belong to the psammomatoid variant.⁷

The presenting symptoms of an orbital JOF will vary according to the size and location of the tumor, from painless bone swelling, globe displacement, to diplopia, and visual impairment due to apical compression. Our

patient noticed proptosis, with no other accompanying symptoms.

Although it is a benign tumor, its growth can cause not only local destruction and facial disfigurement, but also invasion of adjacent vital structures. Thus, complete resection is the treatment of choice.^{1,5,8} With our patient, the decision was made to operate to avoid the development of symptoms of optic nerve compression, since MRI images already showed caudal displacement of the nerve. Radiotherapy is contraindicated.³

Considering the tumor's size and location, approach options were transcranial via the anterior fossa (neurosurgical approach), or transfrontal, either through a bi-coronal incision, or a supraciliary incision, which was the one chosen for our patient. A titanium mesh was placed to restore the left orbital roof, aligned with the right side.

Recurrence after resection has been reported from 30 to 56% which could be related to incomplete excision.^{2,3} No malignant transformation is reported in the literature.² Long-term monitoring is advised, especially in the presence of a psammomatoid histopathology, as is the case of our patient.⁸

CONCLUSION

Juvenile Ossifying fibroma is a rare benign tumor of the craniofacial skeleton, histologically benign but locally aggressive. The orbit is a very rare site of involvement, with risk of potential compromise of the optic nerve. Early diagnosis and thorough treatment are crucial to avoid damage to vital intracranial structures. Surgical removal is the treatment of choice for these tumors. We also highlight the importance of regular patient follow-up because of high recurrence rates.

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REFERENCES

1. Nicolai P, Castelnuovo P. Benign Tumors of the Sinonasal Tract. In: Paul W. Flint PW, Haugh BH, Lund V, Niparko JK, K. Thomas Robbins, Thomas JR, Lesperance MM, editors. *Cumm Otolaryngol Head Neck Surg*. 2015:740-51.
2. Lemoine S, Cassagnau E, Bertin H, Poisson M, Corre P, Guiol J. Juvenile ossifying fibroma: case report and literature review. Management and differential diagnosis. *J Oral Med Oral Surg* 2018;24:67-71.
3. Linhares P, Pires E, Carvalho B, Vaz R. Juvenile psammomatoid ossifying fibroma of the orbit and paranasal sinuses. A case report. *Acta Neurochir* 2011;153:1983-8.
4. Maiorano E, Slootweg PJ. Maxillofacial Skeleton and Teeth. In: Cardesa A, Slootweg PJ, Gale N, Franchi A, editors. *Pathology of the Head and Neck*. 2nd ed. Springer. 2016:179-227.
5. Chrcanovic BR, Gomez RS. Juvenile ossifying fibroma of the jaws and paranasal sinuses: a systematic review of the cases reported in the literature. *Int J Oral Maxillofac Surg* 2020;49:28-37.
6. Cruz A, Alencar V, Figueiredo A, De Paula S, Eichenberger G, Chahud F, et al. Ossifying Fibroma: A rare cause of orbital inflammation. *Ophthal Plast Reconstr Surg*. 2008;24:107-12.
7. Wakefield MJ, Ross AH, Damato EM, Salvi SM, Baker GRC. Review of Lateral Orbital wall Ossifying Fibroma. *Orbit* 2010;29(6):317-20.
8. Lyons CJ. Other mesenchymal abnormalities. In: Lambert S, Lyons C. *Taylor and Hoyt's Pediatric Ophthalmology and Strabismus*. Elsevier Inc. 2017:2328

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