



Oftalmología en imágenes / **Ophthalmology in images**

The importance of the study of proptosis in children

La importancia del estudio de la proptosis en niños

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A 15-year-old boy presented with history of a previous orbitotomy due to an ethmoidal osteoma on his right side, painless progressive protrusion of the right eye (RE) for 6 months. No history of trauma. Normal development and no past family history.

His visual acuity was RE 20/80 and LE 20/20. Pupils were reactive in both eyes. Right proptosis of 25 mm with restriction of the ocular motility superiorly, inferiorly, laterally and medially. Mild conjunctival chemosis and lateral eye displacement (Figure 1A)

Laboratory investigations showed an erythrocyte sedimentation rate of 50 mm/h, the rest of the tests were normal

An MRI was done, finding the following: heterogeneously enhancing of the mass in the right retro-orbital space pushing the eyeball anteriorly and displaced the optic nerve laterally. Sclerosis of the adjacent ethmoid bone but no frank destruction was seen. No extension of the mass into intracranial cavity. Left eye was normal. (Figure 1B)

The presumptive diagnosis was tumor recurrence of primary Osteoma vs Rhabdomyosarcoma in the RE.

Given the clinical and imaging characteristics, the size of the mass and the compromise of the ocular structures, it was decided to perform a biopsy by excision under general anesthesia in the compromised eye.

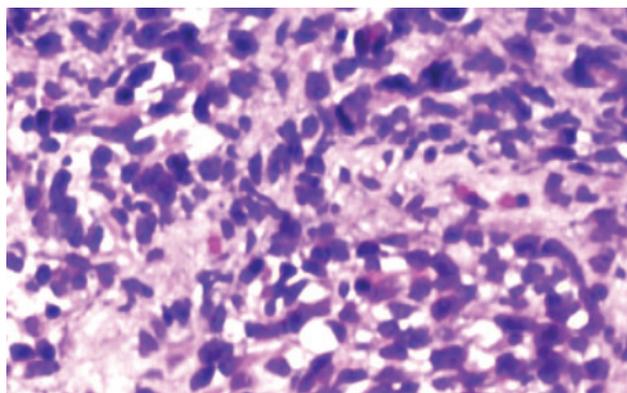
Figure 1: Frontal view of the patient(A) and craneal MRI : Sagittal cut (B)



- A. *Proptosis of the right eye with mild chemosis. The eye is displaced laterally.*
 B. *Low to intermediate intensity mass, hyperintense to the adjacent muscle located in the upper orbit. Retro-orbital mass pushing the eye ball laterally*

The histological diagnosis was an embryonic rhabdomyosarcoma (Figure 2), Using the IRS- post surgical staging system the patient was considered in the group II, so the oncologist started chemotherapy with Vincristine, Actinomycin D and Cyclophosphamide associated with Radioterapy (conventional fractional irradiation). Chest-CT, TC bone scan, and bone marrow punctures and trephines were ordered. Patient maintains treatment to date with periodic follow-ups.

Figure 2. Histology sample



hyperchromatic nuclei, rounded or slightly ovoid, with occasional mitosis, poor cytoplasm and strongly eosinophilic.

Orbital Rhabdomyosarcoma should be considered in the differential diagnosis of any child with a progressive unilateral proptosis. It is also very important to make an adequate classification of the tumor stage in order to start the appropriate treatment. After the biopsy, staging for orbital Rhabdomyosarcoma should be always done according to the IRS post-surgical staging system.¹

This patient was in the stage II. Therefore, chemotherapy and radiotherapy was started.

Some author agree that this classification is useful in terms of treatment, stratification and prognosis prediction.²

The pictures are taken and published with authorization of parents, and informed consent was signed.

REFERENCES

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