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Retro-orbital mature teratoma: a case report *and review of the literature*

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ABSTRACT

Background: We report a case of a 2-month-old female infant with a right retro orbital mature teratoma. Patient operates at two months age old by right lateral orbital approach benefited of a complete resection. Outcomes were aesthetically favorable but functionally unsure.

Key words: Teratoma, retro-orbital, congenital tumor, exophthalmos

Introduction

The retro orbital teratoma is a congenital tumor that is often benign mature and extremely rare. Indeed, since its first description in1862 to date, fewer than 80 cases have been reported.
It is a tumor formed from germ cells that usually affects female infants.
Clinically, the teratoma is characterized at birth by unilateral, axial, and sometimes massive exophthalmos, leading to deformity of the face.
Given the rarity of this type of tumor we report a single case study of a mature teratoma of the right orbit in a two-month-old infant.

Observations

The infant named HH is 2-month-old female born of a non-consanguineous marriage.
The pregnancy was well supplemented, well followed antenatally, and the infant was born by uncomplicated normal vaginal delivery.
HH was admitted to the neurosurgery department of Blida University Hospital for the management of a right retro orbital mass process.

The History of the disease

The pathology was first noted in the delivery room post-natally within the first hours after birth.
The clinical finding of a right exophthalmos was the indication for referral.
Neuroradiological exploration was then carried out on day 4 post-natally, the results of which were clearly indicative of a teratomatous lesion.

The clinical examination signs

Unilateral axial right exophthalmos which was irreducible, non-pulsatile, and globular.
Distension of the eyelids with repressing of the globe, accompanied by conjunctival keratinization, exposure keratinization and corneal ulceration.
We also noted damage of the right extrinsic and intrinsic ocular muscles. (Figure 1)

Cerebro-orbital MRI

The MRI showed a right intra-orbital mass process without signs of locoregional invasion, no anomalies of the optical pathways, and right exophthalmos of grade III. (Figure 2)

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Abdominal ultrasound
Was normal.
The alpha-fetoprotein level was elevated at 104.43 IU / ml (normal level <20 IU / ml).
HH operated at two months age old by right lateral orbital approach benefited of a complete resection.
Macroscopically the piece was globular, its outer face was lobulated. (Figure 3)

The anatomopathological exam
The results were in favor of a mature retro orbital teratom**.**

Postoperative evolution
Post-operative outcomes
Were satisfactory, marked by the exophthalmos' regression, straight convergence of the right eye, and persistence of the right oculomotricity damage. (Figure 4)

The anatomopathological exam
The results confirmed a mature retro orbital teratoma. (Figure 5)

Discussion
Teratomas are impressive but rare congenital tumors. Described for the first time by HOLMST in 1863 [1], their prevalence remains very low. They are usually diagnosed post-natally, however, some reported cases have been diagnosed in the prenatal period [2].
This mass process is similar to a mandarin that distends the eyelids without knowing very well the fate of the eyeball.
Teratomas are congenital multi-cystic masses that occur most often in the gonads but may occur on other sites whose orbit [3, 4].
Teratomas are more prevalent in infant girls, involve the right orbit more often than the left, and are usually not associated with other birth defects or genetic syndromes[5, 6].
Histologically, teratomas are tumors composed of a wide variety of tissues which are foreign to the anatomical site where the teratomas occur (cartilaginous tissues, pilosebaceous appendages calcifications, etc...) [7].
Exceptionally, they may contain parts of the fetal body, or even result in a complete embryo [8].
These are rare congenital tumors with embryonic germ cells. These tumors are characterized by the presence of ectodermal, mesodermal and endodermal components, they may consist of two-cell dermal tumors germ [6]. Malignant transformation is possible but remains extremely rare [9, 10, 11] and may even generate metastases [12],. Clinically they usually present in a newborn with exophthalmos with marked stretching of the eyelids, and elongation of the palpebral fissure, the tumor mass being transilluminated on each side [4]. The large intra-conical masses may be associated with massive exophthalmos accompanied by conjunctival keratinization, exposure keratopathy and ulceration of the cornea [4]. The optic nerve may be embedded or adherent to the tumor resulting in optic atrophy and poor pupillary reaction [4, 13, 14].

The contribution of medical imaging
On the CT scan the teratoma appears irregular, describing a heterogeneous mass with fleshy and cystic components, internal calcifications may indicate the presence of bones or teeth. When the contrast product is injected the fleshy part will be slightly raised. MRI shows a lesion of fleshy and cystic components at sequence T1.
The presence of intracavitary fat and at the level of the walls of the cyst leads to an intense image in sequence T1 and hypo-intense in T2.
The injection of gadolinium moderately enhances the fleshy part.
The differential diagnosis is of rhabdomyosarcoma - normally occurring around the age of 6 to 7 years-, dermoid cyst, cephalocele, lymphangioma and hemangioma. The treatment always consists of the widest curative excision possible, with exenteration of the orbit. It should be noted that this has resulted in good postoperative results with no recurrences (15).
As treatment is aimed at being curative, the eye can not be saved [16].

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As treatment is aimed at being curative, the eye can not be saved (16)
However, it is now possible, thanks to excision and preservation of the eye, to obtain excellent functional and cosmetic results [11, 17, 18].

Conclusion

The retro orbital teratoma is a very rare tumor of very precocious discovery.
Indeed, the diagnosis is made at the neonatal age.
The excision remains macroscopically total, outcomes aesthetically favorable but functionally unsure.

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Figure 1: Unilateral axial right exophthalmos

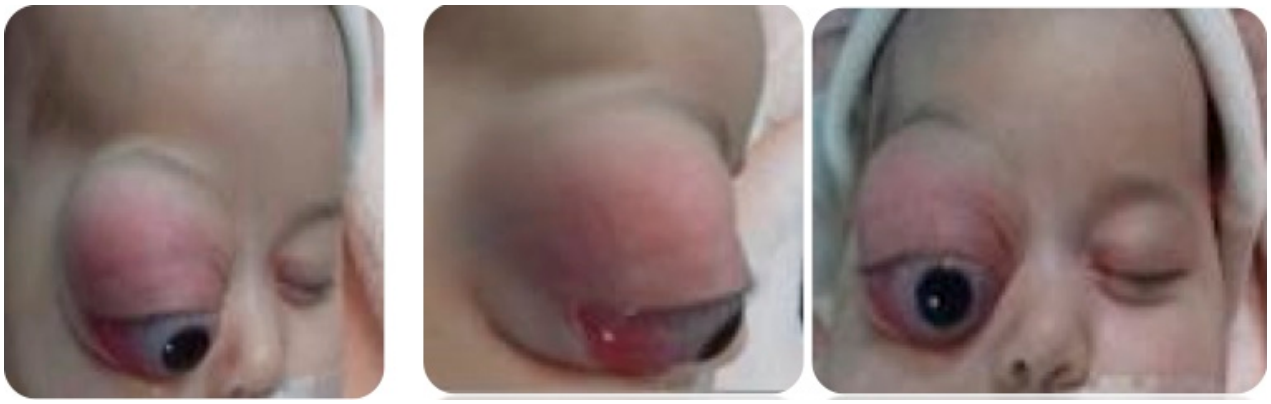


Figure 2: Cerebro-orbital MRI
Right intra-orbital mass process without signs of locoregional invasion, no anomalies of the optical pathways, and right exophthalmos of grade III.

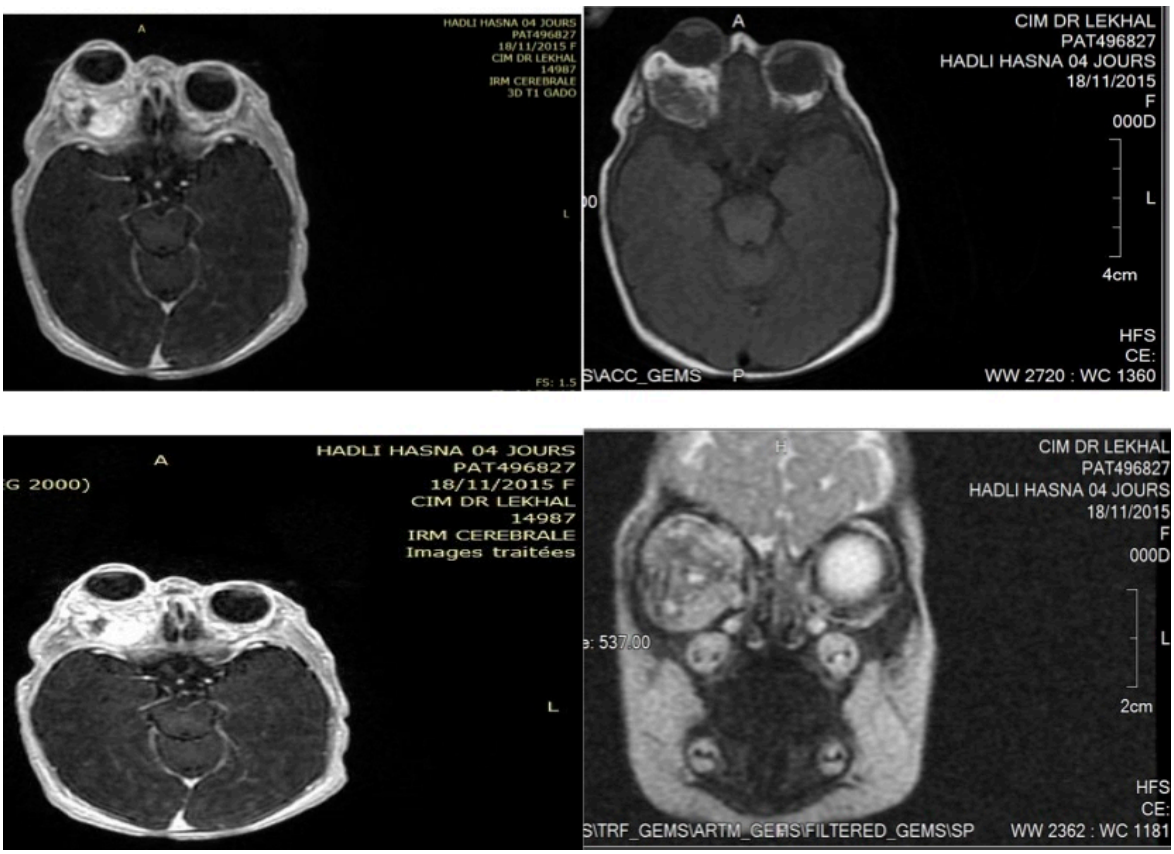


Figure 3: Surgical piece

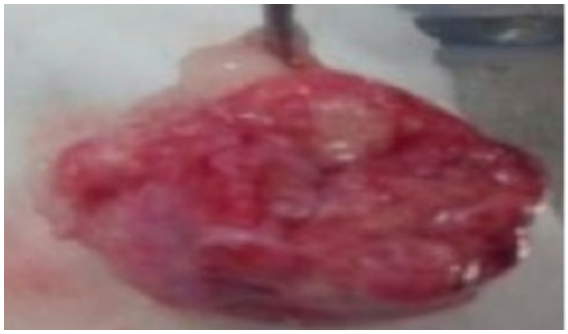
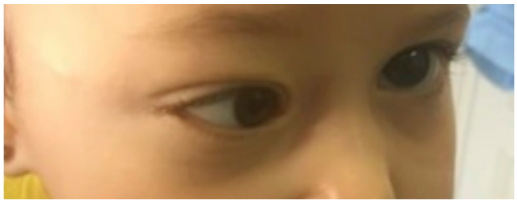


Figure 4: Post-operative results at month 13



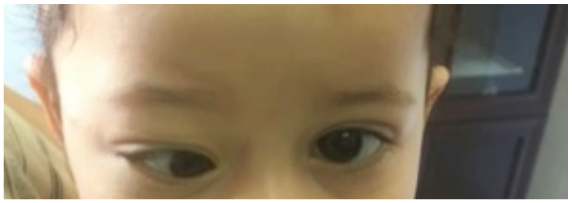
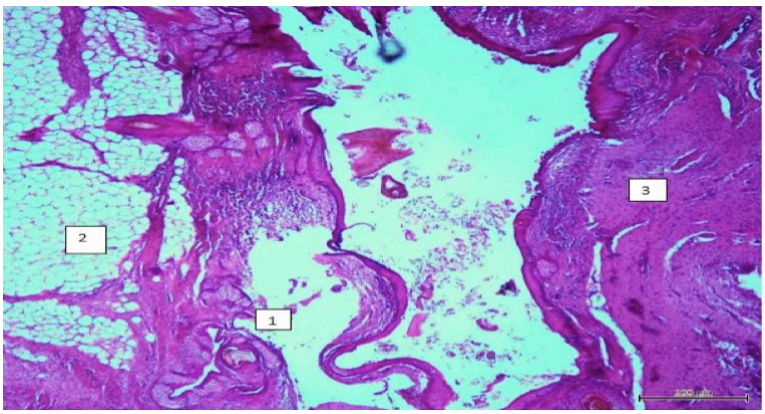


Figure 5: Teratoma mature pluritisular: dermoid cyst, epidermal cyst with sebaceous glands), adipose tissue and mature glial



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