Short communication

Congenital proptosis secondary to orbital teratoma. Clinicopathological study


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A R T I C L E  I N F O

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A B S T R A C T

Case report: A newborn male with right proptosis secondary to a retroocular mass was admitted. Ophthalmological examination also showed corneal ulcer and perforation, iris hernia, total ophthalmoplegia, chemosis and eyelid retraction. The histopathology diagnosis was mature teratoma.

Discussion: Teratomas are tumors composed of a mixture of mature tissues consisting of 3 germ layers. Congenital teratomas of the orbit are very rare and should be included as a possibility in cases with a primary tumor in the orbit.

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Proptosis congénita secundaria a teratoma orbitario. Estudio clinicopatológico

R E S U M E N

Caso clínico: Recién nacido masculino con proptosis secundaria a tumor retroorbitario. La exploración oftalmológica mostró úlcera corneal, perforación, hernia del iris, oftalmoplejía y retracción palpebral. Se realizó estudio anatomopatológico con diagnóstico de teratoma maduro.

Discusión: El teratoma congénito maduro es una neoplasia germinal con presencia de elementos maduros de las tres capas germinales. Es una lesión poco frecuente en la órbita que debe ser incluida en los diagnósticos diferenciales cuando se encuentra un tumor retroorbitario congénito.

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Introduction

Congenital orbital teratoma is an infrequent tumor that originates in the germinal cells which have escaped the influence of organization in early embryo development. The most frequent location of teratomas is the gonada, although they can also be observed in the sacrococcygeal and retroperitoneal region. Mature teratomas are tumors comprising a combination of mature tissue which are characteristic of the 3 germinal layers. The tumors that only exhibit elements of one or 2 germinal layers are choristomas or teratoids tumors respectively.

Case report

A newborn male with right proptosis secondary to retro-orbital tumor was admitted. Ophthalmological physical exploration revealed perforated corneal ulcer, iris hernia, complete ophthalmoplegia, chemosis and palpebral retraction (Fig. 1). Computerized tomography revealed a heterogeneous retro-orbital tumor with cystic and solid areas limited to the orbit with bone reshaping (Figs. 2 and 3). Complete extraction of the tumor was performed with ocular globe enucleation without peri-surgery complications. The tumor was sent for histopathological study.

Macroscopically, the tumor was ovoid and of yellowish coffee color with a size of 4.7 cm × 3 cm × 3 cm. Serial sections revealed a heterogeneous and predominantly solid shiny yellow surface with fatty appearance in addition to small, condroid-looking pearly white areas. In addition, the tumor exhibited small clayish cysts with greenish-yellowish mucus and white lumpy material (Fig. 4). Numerous histological sections were made which in the solid areas exhibited fatty tissue, bone, cartilage, neuroepithelium areas with ganglion cells and choroidal plexus (Fig. 5). The cystic areas were covered with keratinizing stratified flat epithelium similar to epidermis as well as ciliated pseudostratified column epithelium producing respiratory-type mucus (Fig. 6). No immature or malign tissue was observed. The histopathological diagnostic was mature teratoma.

After 18 months in follow-up the patient did not exhibit recurrence of the lesion.
Discussion

Congenital orbit teratomas are rare lesions. Bibliographic reviews refer a slight predominance of the left orbit (60%) and a ratio of 2 to 1 of female patients. Typically, the tumor presents with rapid growth, unilateral proptosis and palpebral retraction without intracranial involvement. Generally, the ocular globe is displaced forward and exhibits degenerative changes secondary to the tumor. In these cases, the orbit exhibits an increase of up to 3 times its size.\(^3\)

At the histological level, congenital teratomas are predominantly benign with mature tissue originated in the ectoderm (flat stratified epithelium, cutaneous annexes), neuroectoderm (glyal tissue, choroidal plexus, ganglion cells), mesoderm (smooth muscle, fatty tissue, bone and cartilage) and less
frequently the endoderm (respiratory and gastrointestinal epithelium).²,⁴

Very few malignant teratoma cases have been published and most correspond to mixed germinal tumors (teratoma with seminoma, embryo carcinoma or endoderm sinus tumor), teratomas with non-germinal malignant tumors (sarcoma, neuroblastoma) and immature teratomas.⁵ The treatment of choice is tumor resection with ocular globe exenteration and preservation of eyelids. However, when the ocular globe is intact conservative surgery should be carried out.²

The present case is a newborn male which exhibited a very rare congenital tumor located in the right orbit. For this reason the authors deemed it important to report it. In the presence of a retro-orbital congenital tumor teratoma must be taken into account in the differential diagnosis. Even though this case did not exhibit malignant or immature elements, the histological study must include multiple histological sections of the tumor for adequate study and classification.

Conflict of interests

No conflict of interest has been declared by the authors.

REFERENCES