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Ciliary Body Tumor in Children: Diagnostic and Therapeutic Problems

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Abstract:
Ciliary body tumors in children present problems of etiological diagnosis. Imaging is essential but confirmation is based on pathological examination which allows the histological nature to be clarified. In our article, we report the case of a tumor of the ciliary body in a girl aged 2 years and 2 months, presenting a tumor of the ciliary body of the left eye revealed by exotropia and a cataract.
Keywords: *tumor - ciliary body – child*

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I. Introduction

The diagnosis of a ciliary body tumor in children can be difficult despite advances in imaging systems, due to the retro-iris situation and the multitude of differential diagnoses. Confirmation of the diagnosis is anatomopathological. It remains essential to offer rapid and adapted treatment to each patient.

II. Case report

The case concerned a 2-year and 2-month old girl, N.H., admitted for treatment of a left unilateral congenital cataract. Having a history of first degree consanguinity and father treated for colorectal cancer, declared cured. The history of the disease dates back to birth with the observation by the parents of a bad red reflex in the left eye associated with tearing. The evolution was marked by poor visual behavior and leukocoria in the same eye. Ophthalmological examination found exotropia in the left eye, positive menace, dazzle and pursuit reflexes, temporal episcleral telangiectasias (Figure 1), a clear cornea, and a hypothalamic anterior chamber (Figure 2), posterior synechiae (Figure 3), an intumescent white total cataract, tone is normal on bi-digital palpation, fundus examination is inaccessible. Examination of the right eye was normal.

Figure 1: temporal episcleral telangiectasias

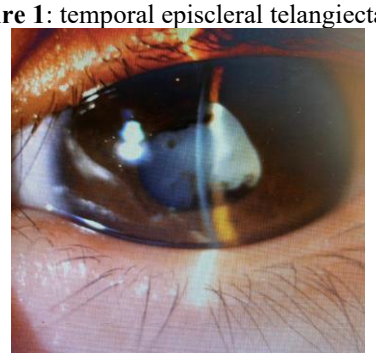

Figure 2: hypothalamia of the anterior chamber

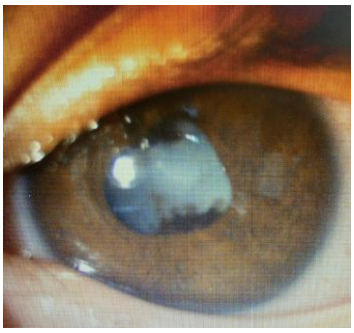

Figure 3: posterior synechiae

Skin examination of the face reveals a hemangioma of the right hemiface which extends towards the right temporal region and the nape of the neck (Figure 4) and erythematous plaques in the left cheek region (Figure 5).

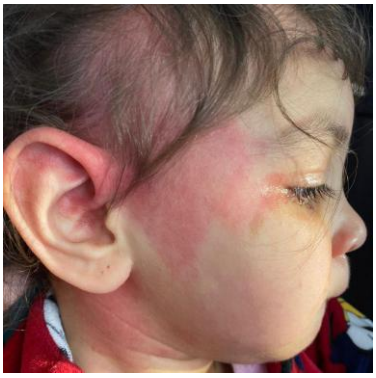

Figure 4: hemangioma of the right side of the face extending towards the right temporal region and the nape of the neck

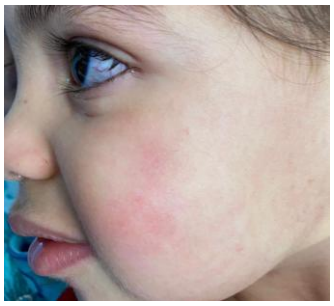

Figure 5: erythematous plaques in the left cheek region

Ocular ultrasound showed the presence of a heterogeneous upper nasal dome formation measuring 7.6 mm/4.53 mm without calcification (Figure 6).

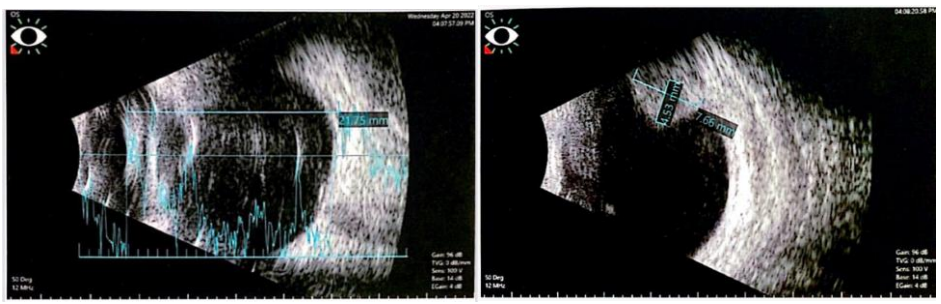

Figure 6: Ocular ultrasound showing fine intravitreal echoes and a heterogeneous dome-shaped lesion in the superonasal region, without calcifications within it.

The cranio-orbital MRI showed a left intraocular expansive process involving its anterior and lateral segment, in T1 iso-signal, moderate T2-diffusion hypersignal with a low ADC, enhanced after injection measuring 5.6x10.3 mm , it is associated with a hemorrhagic lateral retinal detachment, without extension to the conical fat revealing firstly a retinoblastoma (Figure 7).

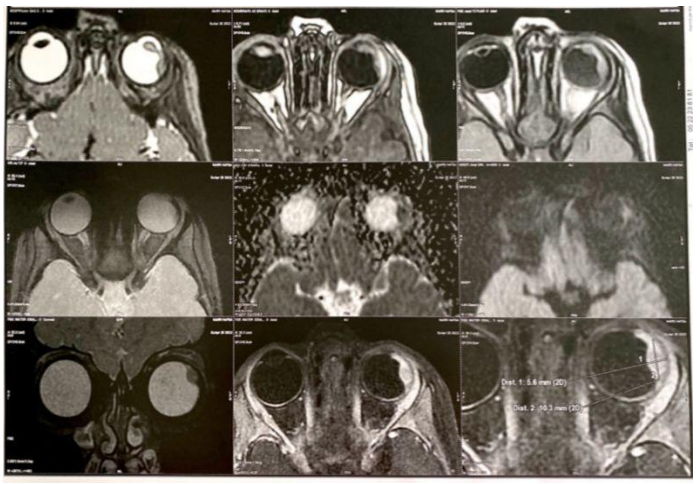

Figure 7: MRI images showing a left intraocular space-occupying lesion involving the anterior and lateral segments, with iso-signal intensity on T1-weighted images and moderate hyperintensity on T2-weighted diffusion sequences with low ADC values. The lesion enhances after contrast injection and measures 5.6 × 10.3 mm. It is associated with a lateral hemorrhagic retinal detachment.

A second control ocular ultrasound was performed showing an increase in the size of the tumor: 10mm/6.6mm versus 7.6mm/4.53mm (Figure 8).

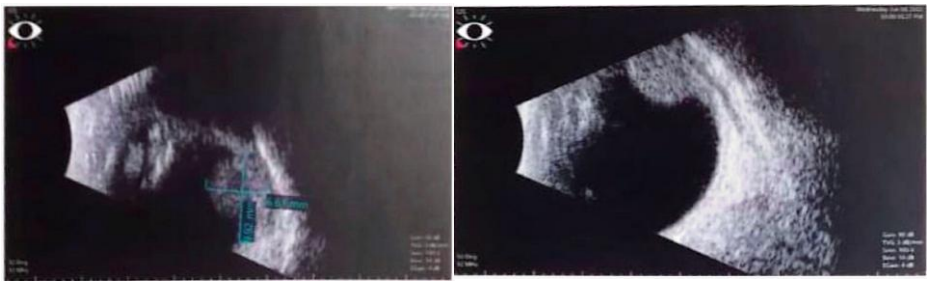

Figure 8: Ocular ultrasound showing an increase in tumor size: 10 mm × 6.6 mm versus 7.6 mm × 4.53 mm.

The UBM of the left eye showed formation most likely at the expense of the ciliary body in the temporal region (Figure 9).

Figure 9: UBM showing a lesion most likely arising from the ciliary body in the temporal region of the left eye.

A dermatologist's advice was requested in view of the facial hemangioma and the “café au lait” skin spots which came back in favor of a suspicion of neurofibromatosis type 1. A brain scan and an abdominal ultrasound were done which returned without particularities. The patient underwent cataract surgery with tumor biopsy and cryotherapy next to the tumor. The anatomopathological examination concluded that there were no signs of malignancy. An ocular ultrasound was performed postoperatively which showed disappearance of the tumor, fine intravitreal echoes and a flat retina.

III. Discussion

The ciliary body is the intermediate segment of the uvea. It has a double embryological origin: mesodermal, giving the ciliary muscle, melanocytes, stroma and vascular endothelium, and ectodermal giving the non-pigmented and pigmented epithelia. This large tissue variety explains the variety of different benign or malignant tumors that can affect the ciliary body. We find pigmented lesions such as melanocytoma [1] and ciliary body melanoma [2], epithelial adenoma [3] (or rarely adenocarcinoma [4]), metastases [5], benign lesions such as schwannoma [6], leiomyoma [7], rare diseases such as hemangiopericytoma [8], medulloepithelioma [9, 10] or rarely inflammation such as giant sarcoid tumors [11]. A diagnostic biopsy is usually necessary. Uveal melanocytoma is a benign tumor that can be diagnosed at any age. Cases have been documented in the iris, ciliary body and choroid [12-13]. Ciliary body melanocytoma is usually hidden behind the iris and is only recognized when it is more advanced. Most are managed by iridocyclectomy. The main complications are cataract and secondary glaucoma. Uveal melanoma is a primary intraocular malignancy that can occur at any age, even as a congenital tumor. Neurofibromatosis type 1 could play a role in its development [14]. Early signs include dilation of the episcleral vessels (or "sentinel vessels") and low intraocular pressure compared to the healthy eye, 5 mm Hg or more. Treatment is often based on radiotherapy. Ciliary body medulloepithelioma is a rare congenital intraocular tumor [15], which often occurs between the ages of 2 and 5 years [16]. It usually presents with leukocoria and a mass arising from the ciliary body. Ultrasound biomicroscopy helps guide the diagnosis. The positive diagnosis is based on histological and immunohistochemical studies. There is no standardized treatment; enucleation remains the most common treatment in the absence of extra-scleral extension [17]. Ciliary body adenoma is a rare benign tumor and very often initially diagnosed as melanoma [18]. We often find the presence of a nearby cataract or a dislocation of the lens. There are no sentinel vessels as in a ciliary body melanoma. Only histology allows the diagnosis to be made [19]. Fine aspiration or local resection, often possible by iridocyclectomy, allows the diagnosis to be established. The malignant form or adenocarcinoma is rarer. No metastasis has been described [20, 21]. Leiomyoma is a rare, benign tumor arising from the smooth muscles of the ciliary body. Only 80 cases are described in the literature [22]. It is frequently a discovery following enucleation or iridocyclectomy for melanoma. Leiomyoma is most often located in the ciliary processes in 73% of cases. The lesion is discovered in children or young adults. In our case, given the absence of signs of malignancy on the anatomopathological study and the disappearance of the tumor after the biopsy followed by cryotherapy, the diagnosis of benign tumor remains the most mentioned, however the certainty diagnosis has remained difficult, despite advances in imaging.

IV. Conclusion

Ciliary body tumors in children can pose a problem of diagnostic delay due to their retro-iris location, and can also pose a problem of etiological diagnosis like our case, due to the multitude of differential diagnoses.

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