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Rubeose irienne revealing a retinoblastome: a case report

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ABSTRACT

Retinoblastoma, the most common malignant ocular tumor in infancy, primarily affects children under five, with a peak onset at one year for hereditary bilateral cases and at two years for sporadic unilateral cases. Rarely, it affects older children and adults, complicating diagnosis.

Keywords : *Retinoblastoma; Malignant ocular tumor; Childhood eye cancer; Strabismus; Unilateral retinoblastoma; Endophytic retinoblastoma;*

1. INTRODUCTION

Retinoblastoma is the most common malignant ocular tumor in infancy, affecting children under the age of five. In hereditary bilateral cases, the peak age of onset is one year, while in sporadic unilateral cases, it is two years. Although rare, the tumor can also affect older children and adults, which presents a challenge in differential diagnosis.

2. CASE REPORT

We encountered a 6-year-old child from a first-degree consanguineous marriage who visited our clinic a year ago due to difficulties at school and visual problems related to strabismus. The child had previously received treatment for amblyopia (occlusion), but there was no improvement in visual acuity.

Three months prior to our encounter, the same child, with no significant medical history, sought our consultation again due to difficulties at school. Upon ophthalmological examination of the left eye, we observed positive visual acuity, an ocular tone of 32 mm Hg, iridial rubeosis, and a whitish mass pushing the retina backward. These findings led us to suspect endophytic retinoblastoma. Examination of the right eye revealed a visual acuity of 10/10, with a normal anterior segment and fundus.

Based on these findings, we diagnosed the child with unilateral retinoblastoma in the left eye. The tumor was classified as group E according to the International Classification of Intraocular Retinoblastoma, necessitating chemo-reduction followed by enucleation of the left eye. To restore the eye's appearance, a hydroxyapatite intra-orbital implant was inserted. Pathological examination confirmed the diagnosis, revealing a moderately differentiated retinoblastoma with moderate necrosis. No vascular emboli or extensions into the choroidal or scleral regions were observed. The cribriform lamina showed tumoral involvement, but the resected optic nerve margin was healthy.

Due to the macroscopic involvement of the anterior segment, tumoral infiltration of the ciliary lamina, and involvement of the ciliary processes, adjuvant chemotherapy was recommended. The child is currently being regularly monitored in our department and has been fitted with a prosthesis. In Figure 7, extensive vitreous swarming with a partially obscured papilla can be observed, with a visual acuity of 10/10 in the unaffected eye.

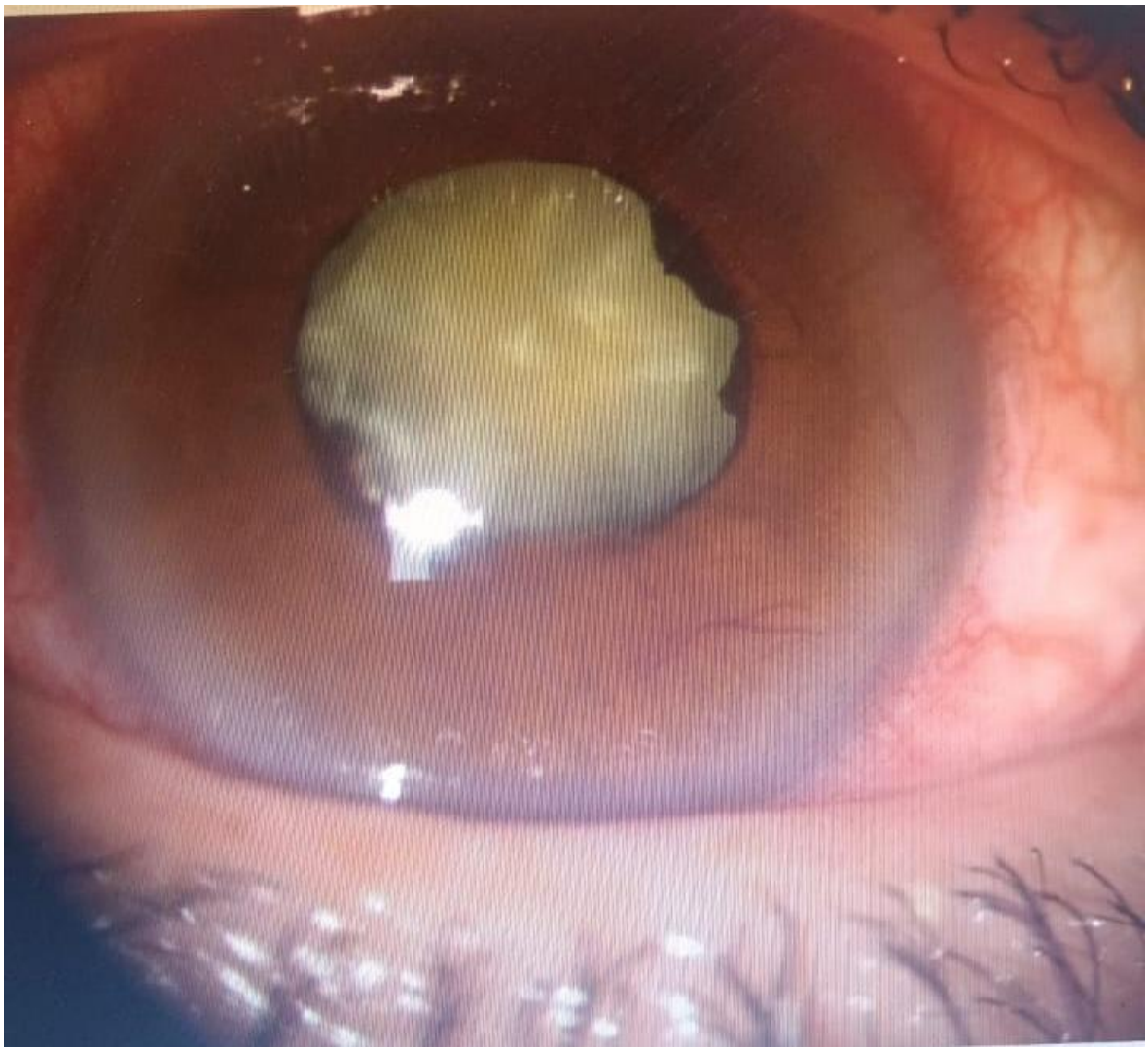

Figure 1: image showing leukocoria with iridal rubeosis

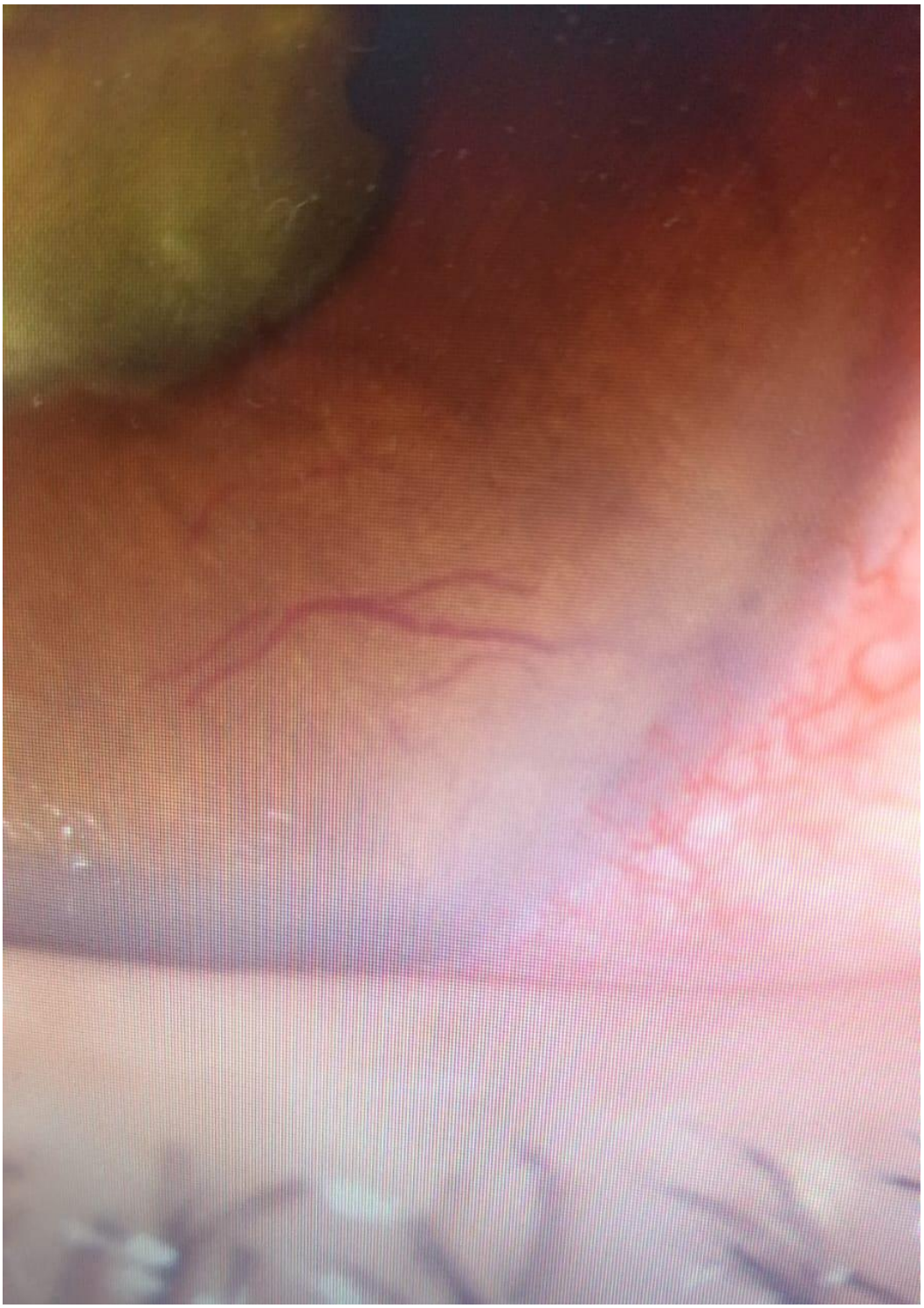

Figure 2: close-up image of iris rubeosis

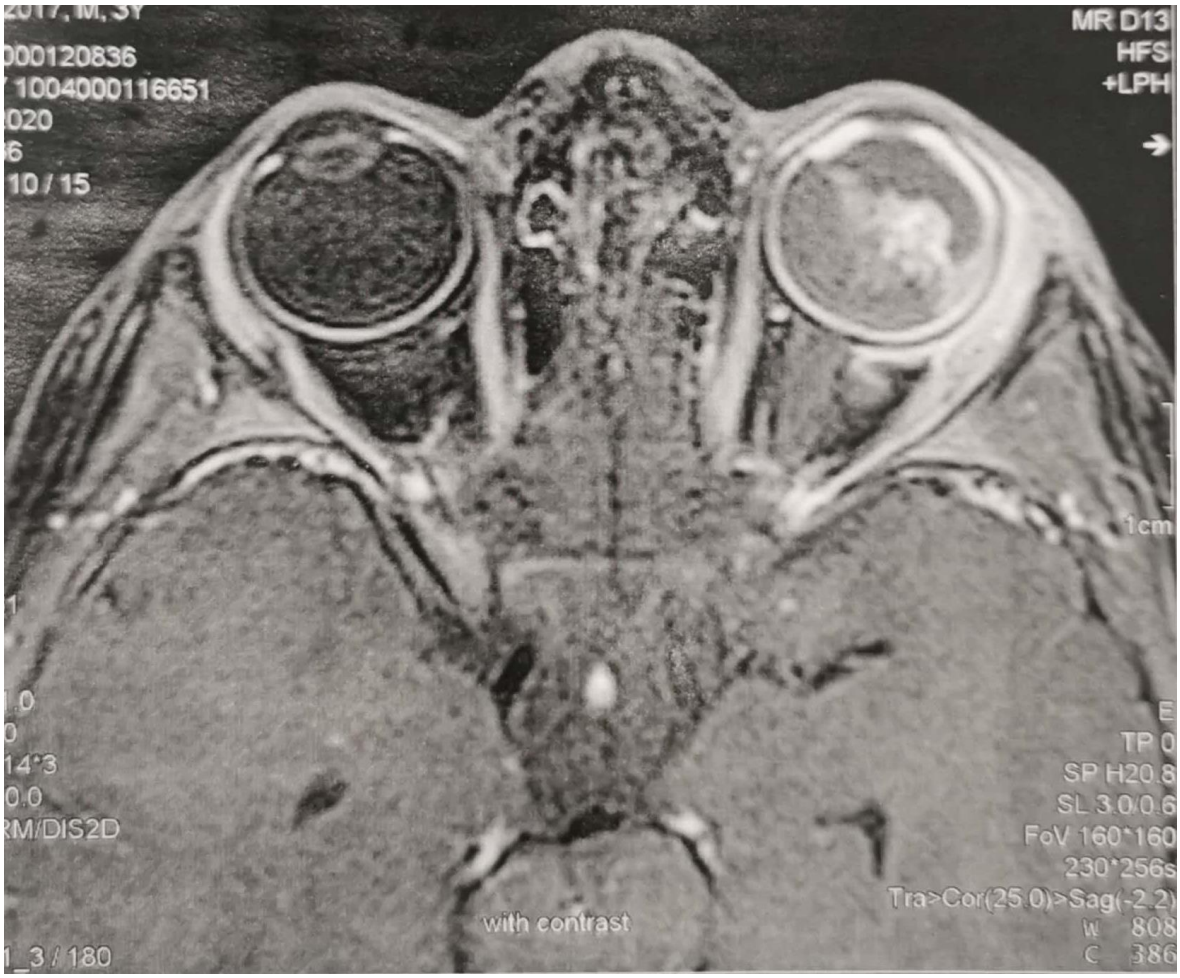

Figure 3: mri showing tumor process suggestive of retinoblastoma

3. DISCUSSON

Retinoblastoma (RB) is a rare malignant tumor that develops in the retina, particularly in children under the age of 5 in 90% of cases. However, cases have been reported in older people, including adults up to the age of 74 [1]. For example, Shields found that around 6.5% of patients were over 5 years of age in a series of 400 patients, and a Tunisian study found that 7% of patients were over 5 years of age [2].

Possible diagnoses included ocular toxocariasis, Coats' disease, and retinoblastoma. Ultrasonographic and CT examinations strongly suggested retinoblastoma, confirmed by an anterior chamber puncture showing increased LDH levels. Enucleation of the left eye confirmed the diagnosis by pathological examination [3].

The fact that RB can occur in older children poses significant problems in terms of diagnosis and treatment. Unlike young children, in whom the main symptoms are leukocoria (abnormal white reflection in the pupil) and strabismus, older children often report a drop in visual acuity. Atypical signs such as hypopyon, uveitis, and neovascular glaucoma can complicate diagnosis, leading to errors and delays in treatment [4].

Imaging techniques such as ultrasound and CT scans play a crucial role in diagnosis. However, in cases of doubt, more invasive examinations, such as anterior chamber puncture, may be necessary to confirm the diagnosis. The presence of calcifications on ultrasonography is a key diagnostic criterion, although atypical forms can sometimes lead to confusion [5].

This case highlights the importance of thorough and accurate diagnostic evaluation in older children with symptoms of retinoblastoma to avoid delays in treatment and optimize clinical outcomes.

4. CONCLUSION

The occurrence of B.R. in an older child poses a challenge in terms of diagnosis, both positive and differential. In addition to occurring at an unusual age, the symptoms are often atypical. Magnetic resonance imaging (MRI) is becoming an increasingly reliable diagnostic tool, helping to reduce diagnostic errors. These errors have serious consequences as they can lead to unnecessary over-ablation and endanger the child's life.

CONFLICTS OF INTEREST

All authors declare that they have no conflicts of interest.

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