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Retinoblastoma Masquerading as Uveitis

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ABSTRACT. The clinical and pathologic findings of a 11-year-old boy with retinoblastoma masqueraded as atypical intraocular inflammation are presented. This clinical diagnosis is difficult. Aqueous aspiration for cytological study is useful and should be evaluated carefully.

Key words : retinoblastoma — pseudohypopyon — cytologic diagnosis

Retinoblastoma usually occurs before the age of 2 years and is rare after the age of 4 years.[1] Although the correct diagnosis is possible in the majority of patients with retinoblastoma associated with clinical characteristics, atypical manifestation occasionally leads to misdiagnosis.

We report the clinical and pathological feature of a retinoblastoma masqueraded as uveitis.

CASE REPORT

A 11-year-old boy was examined because of remarkably decreased visual acuity and inflammation in the left eye on April 18, 1983. He noticed floating spots and blurred vision in his left eye in the first of April 1983.

Examination of the left eye revealed vision with 0.2. The anterior chamber was opaque with keratic precipitates (Fig. 1). Ophthalmoscopic examination revealed the vitreous opacity and bleeding and whitish exudates covering the retina (Fig. 2). The right eye was normal. He received dexamethasone generally and betamethasone drops locally, however, opacity of the anterior chamber and exudates did not decrease.

The chromosome study used G-band method was normal. Ultrasonography showed vitreous debris and a flat elevation of the retina. On April 28, 1983, a paracentesis was performed and cytological examination disclosed neoplastic cells which were consist of small and basophilic cells with hyperchromatic nuclei and scanty cytoplasm (Fig. 3).

The ratio of aqueous lactatic dehydrogenase (LDH) to serum lactatic dehydrogenase was less than 1.0. Isozyme LDH_3 and LDH_4 levels increased in aqueous. Other serological or chemical analysis showed normal levels.

Based on the results of the cytological study, an enucleation was performed on March 6, 1983.

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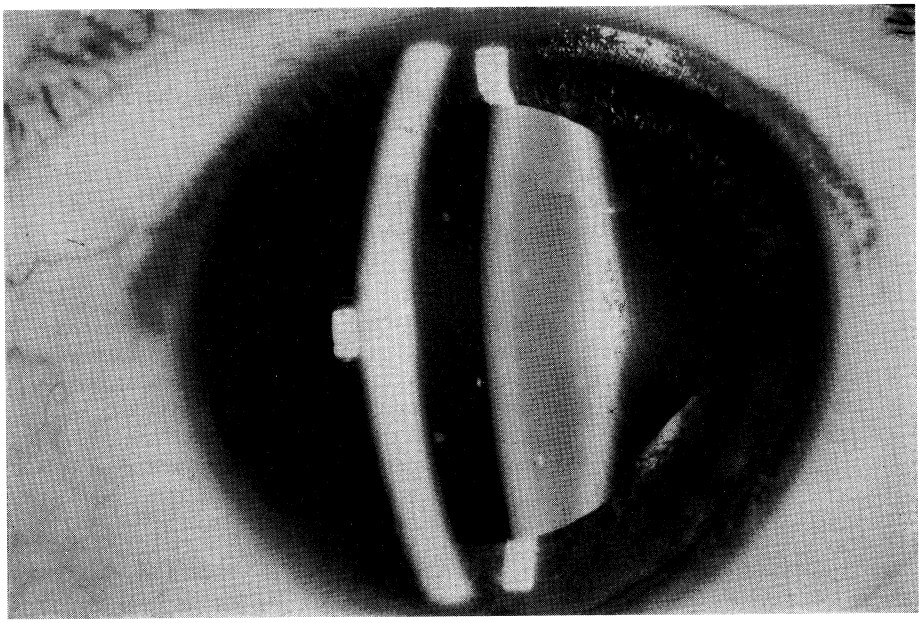

Fig. 1. External photo shows opacity of the anterior chamber with keratic precipitate

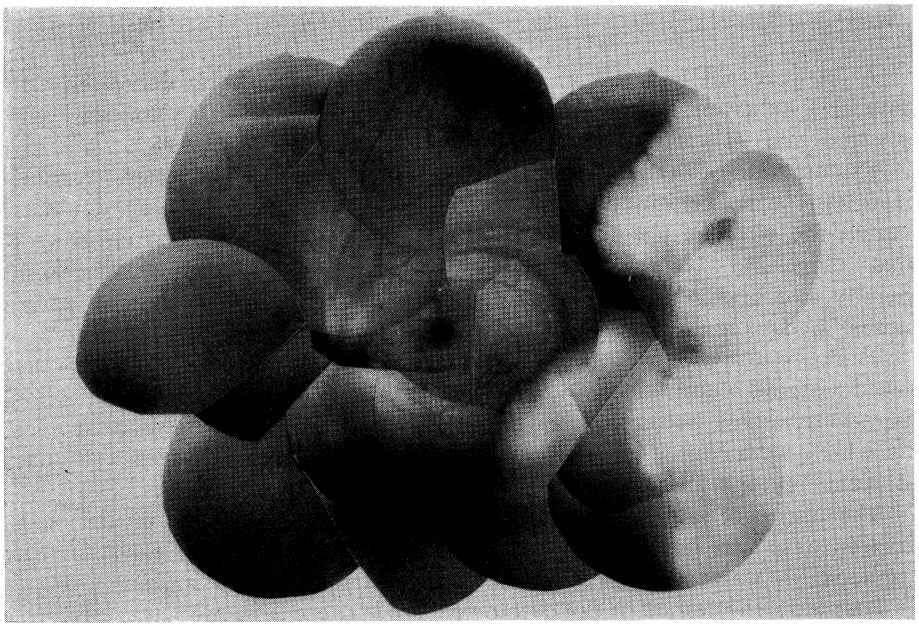

Fig. 2. The left fundus photo reveals a white retinal area in the temporal portion

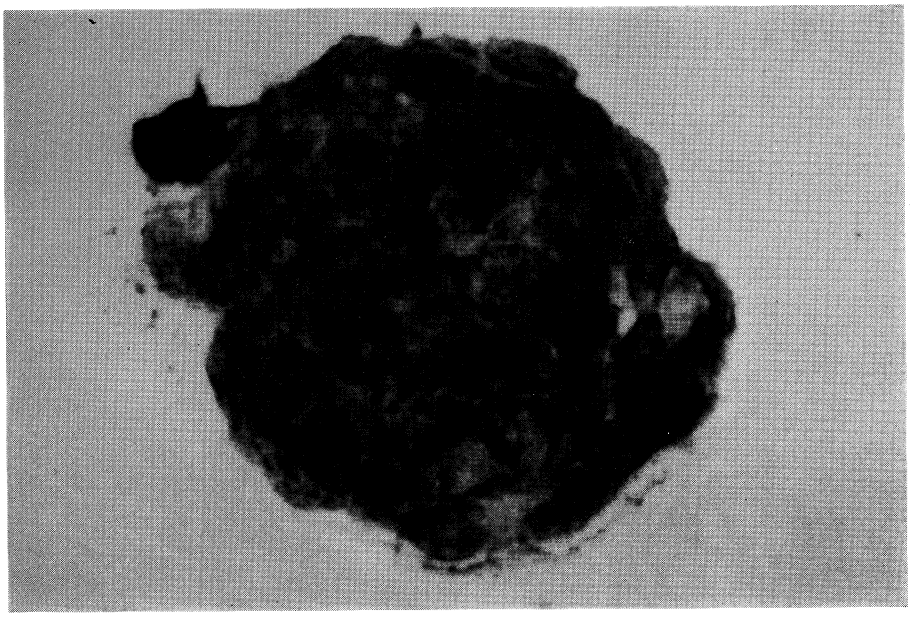

Fig. 3. Aqueous aspirate reveals a cluster of neoplastic cells. H.E. × 200

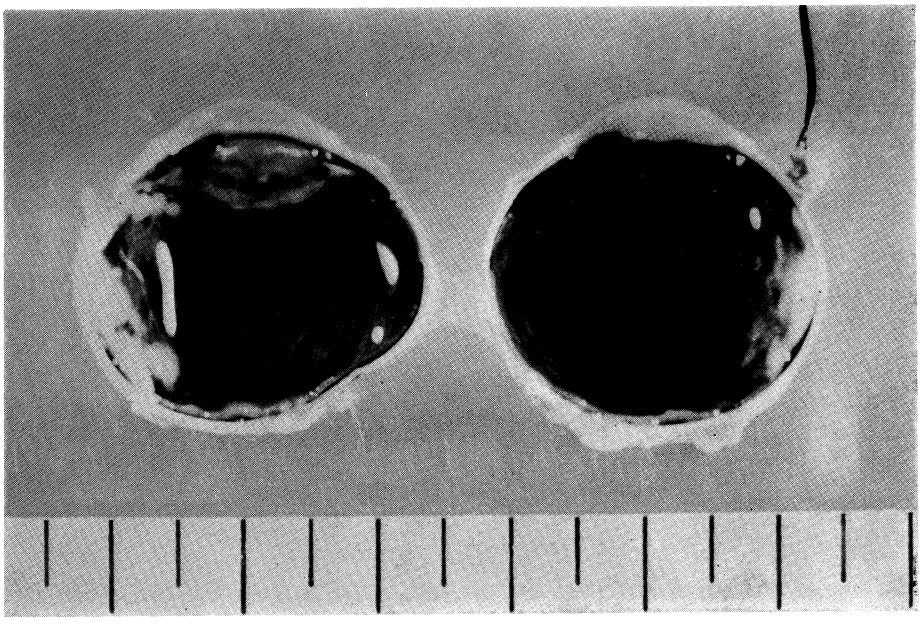

Fig. 4. Grossly the eye shows white tumor in peripheral retina

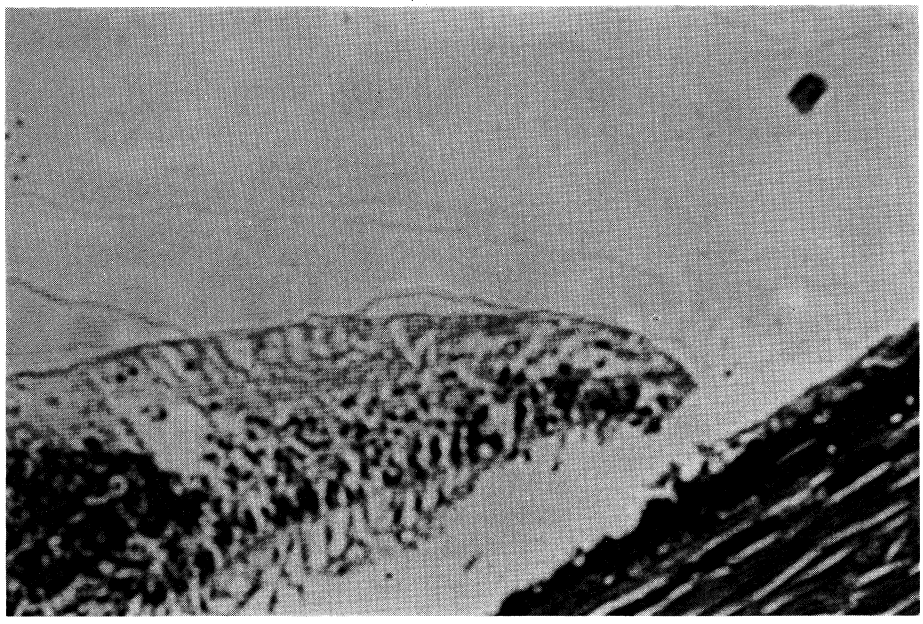

Fig. 5. Tumor arising in the far retinal periphery. V.G. × 50

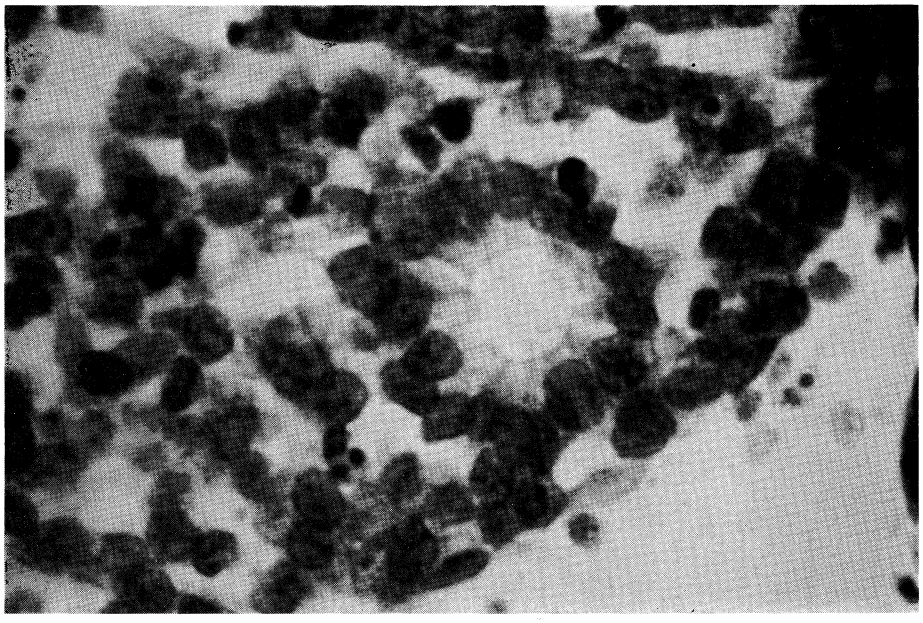

Fig. 6. Tumor cells with rosette formation. Masson : × 500

PATHOLOGY

The globe was opened vertically and showed white soft tumor masses in the temporal region of peripheral retina (Fig. 4).

Histology revealed marked proliferation of small to medium sized round cells arranged in solid fashion (Fig. 5), Necrosis and mitosis exist in the tumor cells remarkably and rosette formation was evident occasionally (Fig. 6).

No invasion of tumor to uvea and sclera was identified. No invasion of tumor cells to optic nerve was present.

DISCUSSION

Retinoblastoma is the most common intraocular tumor in childhood. Bilateral hereditary cases generally have an earlier onset (average 9 months) than unilateral cases of either hereditary or nonhereditary types (average 22 months).[2] This tumor is only a few after the age of 7 years. The diagnosis of retinoblastoma may be difficult, particularly in juvenile and adult patients, because inflammatory signs and glaucoma are the initial manifestations frequently. This type enables the tumor to masquerade as a variety of intraocular inflammatory process.[3-6] In the present case, intraocular tumor, coat's disease, metastatic abscess, nematode endophthalmitis, intraocular inflammation were suspected from the ophthalmoscopic findings.

Zimmerman reports that retinoblastoma is mis-diagnosed in the cases with inflamed eye, history of recent injury, and opacity of media and also in the cases of adult boy. Retinoblastoma is misdiagnosis as uveitis, endophthalmitis, panophthalmitis, scleritis or episcleritis and neuritis.[7]

The present patient was 11 years of age and demonstrated unilateral involvement and a sporadic form of the disease. The family history was negative. His general health was good.

Some cases of retinoblastoma are associated with partial deletion of the long arm of chromosome 13.[8] This case had not abnormalities on chromosome.

Ultrasonography did not show typical finding and a characteristic dampening of tissue echoes posterior to the tumor.[9]

As regarding aqueous humor enzyme levels, Lactate dehydrogenase (LDH) concentration in serum and other extracellular fluids such as aqueous humor is low normally, and the ratio of LDH in aqueous humor to serum LDH concentration is less than 1.0 in control patients with ocular diseases other than retinoblastoma. In contrast, aqueous humor from most eyes with retinoblastoma exhibits increased LDH activity, expressed as an LDH/serum ratio greater than 1.0.[10] In the present case, the ratio of aqueous humor LDH to serum LDH concentration was less than 1.0 but isozyme LDH_3 and LDH_4 levels increased in aqueous.

Aqueous as vitreous aspiration for cytological study may be useful in differentiating retinoblastoma from simulation inflammatory conditions and should be evaluated carefully. Inflammatory cells in the anterior chamber in the eyes with the retinoblastoma may be related to three patterns : (1) diffuse infiltration of the retina without focal retinal mass and extensive seeding of tumor cells into the vitreous cavity and anterior chamber : (2) retinoblastoma

arising in the far retinal periphery with infiltration of anterior segment, and (3) necrotic retinoblastoma leaving scattered neoplastic remnants and inflammatory cells throughout the eye.

Thus, we should note that eyes harboring retinoblastoma may present with clinical signs of ocular inflammation.

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