

Short Communication

RETINOBLASTOMA ASSOCIATED WITH COATS' DISEASE-LIKE FEATURES

The differential diagnosis of leucocoria is well established;⁽¹⁾ however, equivocal ancillary test results may cause confusion between Coats' disease and noncalcifying retinoblastoma with telangiectatic retinal blood vessels.⁽²⁾ Moreover, in Coats' disease, small amounts of calcification may be detected in eyes with predominant exudative and subretinal detachments.⁽³⁾ Here we report findings in a patient with a unilateral retinoblastoma with Coats' disease-like features.

Case Report

A seven-and-a-half-year-old boy presented with a two-month history of a unilateral red eye without antecedent trauma and a negative past medical and family history. Ophthalmic examination revealed a buphthalmic right eye with intraocular pressure up to 45 mmHg. Visual acuity in this eye was reduced to light perception without projection. Fundoscopic evaluation was remarkable for prominent telangiectatic retinal vessels in a detached retina, with yellowish epiretinal and subretinal hemorrhagic exudate surrounding a subretinal mass. The left eye was normal.

Ultrasound assessment revealed a large retinal detachment with exudate, consistent with Coats' disease; however, retinoblastoma could not be excluded as the findings were consistent with the presence of small amounts of calcium. Similarly, the computed tomography scan was interpreted as indicative of Coats' disease, yet the minimal calcification detected could not preclude a suspicion of retinoblastoma.

The painful and virtually blind eye was enucleated for definitive diagnosis.

Histopathological Findings

Grossly, the eye revealed a retinal detachment overlying a mass lesion. Microscopic examination

of stained histopathological sections demonstrated a poorly differentiated and mostly necrotic retinoblastoma involving the detached retina and invading the choroid, the inner scleral layers, and the optic nerve head. The neoplastic cells had rounded to ovoid hyperchromatic nuclei and scanty cytoplasm (Fig. 1). No rosettes were identified. Numerous large, telangiectatic retinal blood vessels were present within the tumor and the adjacent peripheral retina (Fig. 2). The subretinal space contained abundant proteinaceous exudate and scattered tumor cells; however, no cholesterol clefts or lipid-laden macrophages were noted. The anterior segment disclosed rubeosis iridis and peripheral anterior synechiae. The optic nerve head showed posterior bowing of the lamina cribrosa consistent with glaucoma.

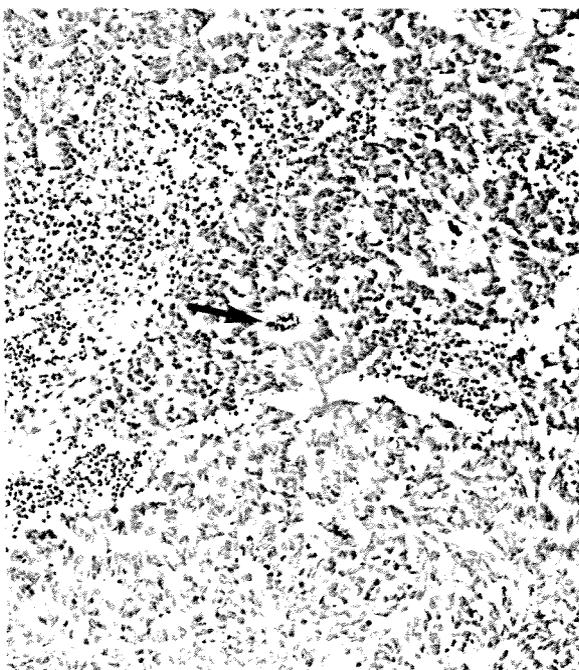


Figure 1. Small blood vessels (arrow) are present within a poorly differentiated retinoblastoma. (Hematoxylin & eosin, original magnification $\times 150$).

Since intravitreal injections of effective antibiotics have been known to fail,⁽⁹⁾ injecting both the posterior and anterior compartments of the eye appeared indicated, although, to my knowledge, this is a novel approach. As the corneal endothelium is far more resistant to the toxic effect of antibiotics than the retina, much higher concentrations may be deposited into the anterior compartments of the eye than may be safely injected into the vitreous cavity. As a consequence, higher concentrations than may be safely injected into the vitreous cavity should diffuse into the region of the plaque overlying the posterior capsule, a region where organisms are presumably present in relative abundance.

Since the volume of the vitreous cavity (4–5 cc) is about 10 to 12 times greater than that of the AC and posterior chamber combined (maximally 0.4 cc), the concentration, in these chambers, of clindamycin must have been approximately 2.5 to 3 times, and that of vancomycin 5 to 6 times as high as that in the vitreous cavity. In view of the shorter half-life of the antibiotics in the anterior chambers of the eye, due to the more rapid turnover of aqueous humor and loss by diffusion into adjacent tissues and the vitreous cavity, use of a higher concentration of these compounds at this site may at times be crucial for achieving a cure.

Conclusion

Delayed onset postoperative intraocular inflammation may be infectious in nature and, if significant, requires anterior chamber and vitreous tap with appropriate microbiological culture techniques to establish the diagnosis. Management is controversial at this time, but intraocular injection(s) of suitable antibiotics, both intravitreally, and intracamerally (where much higher concentrations are tolerated) seem an effective approach. Only when such treatment has failed should more aggressive surgical intervention be considered, including pars plana vitrectomy, partial or total capsulectomy, and removal or exchange of an intraocular lens.

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