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SHORT COMMUNICATION

Sympathetic ophthalmia in VATER association combined with persisting hyperplastic primary vitreous after cyclodestructive procedure

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PURPOSE. *To report on the occurrence of histology-proven sympathetic ophthalmia in a patient with VATER association and persisting hyperplastic primary vitreous (PHPV) after a cyclodestructive procedure was performed to treat secondary angle-closure glaucoma.*
METHODS. *The left eye of a 13-year-old boy with VATER association was microphthalmic from birth and had been diagnosed with PHPV at age 1 year. It developed iris neovascularization and secondary angle-closure glaucoma, which was treated by combined cyclocryocoagulation and cyclophotocoagulation. Six weeks later, a bilateral fibrinous iritis developed. Despite intensive topical and systemic steroid treatment, the iritis persisted so that the left blind eye was enucleated.*
RESULTS. *Histology of the enucleated eye showed a marked intraocular inflammation with lymphocytes, epithelioid cells, and multinuclear giant cells grouped around remnants of melanin-bearing cells.*
CONCLUSIONS. *Sympathetic ophthalmia may occur in patients with VATER association and PHPV after a secondary angle-closure glaucoma is treated by a combined cyclocryocoagulation and cyclophotocoagulation. (Eur J Ophthalmol 2006; 16: 171-2)*

KEY WORDS. *Sympathetic ophthalmia, Persisting hyperplastic primary vitreous, Iritis, Uveitis, Cyclocryocoagulation, Cyclophotocoagulation*

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INTRODUCTION

The VATER associations are characterized by growth failure and pleiotropic congenital malformation (1). The acronym VATER stands for the diagnostic features of vertebral defect, anal atresia, tracheo-esophageal fistulae, esophageal atresia, and radial/renal dysplasia. We describe a young patient with VATER association and persisting hyperplastic primary vitreous (PHPV) who developed sympathetic ophthalmia.

Case report

A 13-year old boy with VATER association with marked growth retardation, vertebral defects, and anal atresia additionally showed in his left eye a unilateral microphthalmos from birth. At age 1 year, it was diagnosed with PHPV (2). At age 13 years, the left eye developed a marked iris neovascularization and secondary angle-closure glaucoma. Due to the intraocular malformations and cataract, the retina could not be visualized upon ophthal-

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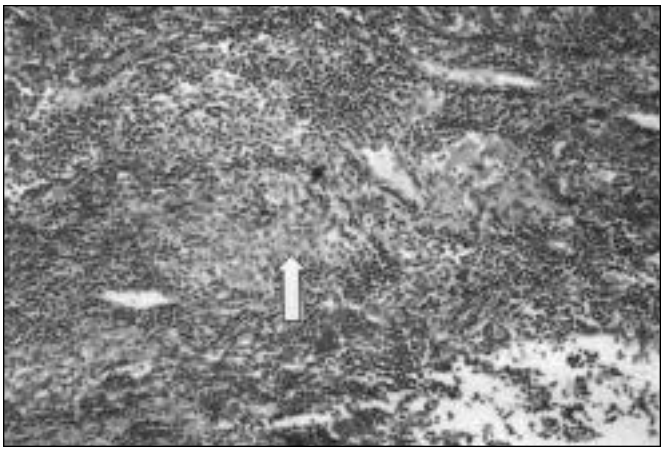

Fig. 1 - *Histophotograph showing a marked intraocular inflammation with multinuclear giant cells (arrow) around melanin-bearing structures.*

moscopy. A combined cyclocryocoagulation and cyclophotocoagulation was carried out. Six weeks later, bilateral fibrinous iritis developed. Despite intensive topical and systemic steroid treatment, the iritis persisted so that the left blind eye was enucleated.

RESULTS

Histology revealed a marked intraocular inflammation with a dense infiltration of lymphocytes, plasma cells, and scattered epithelioid cells and multinuclear giant cells, grouped around melanin-bearing cells and structures representing remnants of the retinal pigment epithelium and iris (Fig. 1). Additionally, calcification was present in the phthisic eye. After enucleation, a combined immunosuppressive treatment of 10 to 15 mg prednisolone, methotrexate, and cyclosporine has resulted in subsiding the remaining iritis in the sympathized right eye. Visual acuity remained at 20/20, and ophthalmoscopy as well as fluorescein angiography were unremarkable. During the follow-up of more than 6 months, reduction of the systemic immunosuppressive therapy has led to a reactivation of the iritis.

DISCUSSION

Sympathetic ophthalmia is a devastating granulomatous inflammation destroying melanin-bearing cells in the posterior region of the eye and leading to bilateral blindness unless systemic immunosuppressive therapy is initiated in the early stage of the disease. Although it was a common reason for bilateral blindness in the wars of the 19th century, when perforating or penetrating ocular injuries were often surgically revised late after the ocular insult or not at all, sympathetic ophthalmia has become a clinical rarity in modern times. Few reports describe the occurrence of sympathetic ophthalmia in patients after cyclodestructive procedures (3-5). Sympathetic ophthalmia has not been reported so far in eyes with the characteristics of a PHPV. Depending on its severity, surgical treatment of PHPV focuses on vision preservation by phakectomy, vitrectomy, or membranectomy to prevent the sequelae of glaucoma and phthisis. The patient described in the present report, however, did not undergo any ocular surgery prior to the development of secondary angle-closure glaucoma, which was treated by cyclocryocoagulation and eventually resulted in sympathetic ophthalmia.

In conclusion, the present report suggests that sympathetic ophthalmia may occur in patients with VATER association and additional PHPV if a cyclodestructive procedure is performed to treat a secondary angle-closure glaucoma.

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