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SHORT COMMUNICATION

Retinocytoma associated with calcified vitreous deposits

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PURPOSE. *To report a case of bilateral retinocytoma associated with calcified vitreous deposits.*
METHODS. *Case report.*
RESULTS. *On routine examination, a 35-year-old asymptomatic father of a child with bilateral retinoblastoma presented bilateral retinocytoma associated with vitreous calcifications, in the vicinity of the retinocytoma in his left eye. Fundus photographic documentation and fluorescein angiography were performed. The patient has been followed up for 10 years.*
CONCLUSIONS. *The lesions in both eyes have remained stable without signs of growth or malignant transformation. Calcified vitreous deposits are a recently described feature of retinocytoma in addition to the three classic features: translucent retinal mass, retinal pigment epithelial alteration, and calcification. (Eur J Ophthalmol 2006; 16: 349-51)*

KEY WORDS. *Retinocytoma, Vitreous deposits, Classic features*

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INTRODUCTION

The presence of a translucent retinal mass, calcification, and retinal pigment epithelial alterations are the three ophthalmoscopic features characteristic of retinocytoma.

Calcified vitreous seeding in the vicinity of the retinocytoma is a recently described feature (1-3).

We report a 35-year-old asymptomatic father of a child with bilateral retinoblastoma who on routine examination presented a retinocytoma associated with diffuse vitreous calcifications in the left eye.

Case report

In 1993, the 35-year-old father of a 1-year-old child with bilateral retinoblastoma underwent ophthalmologic examination, as routinely performed on relatives of children with retinoblastoma. The patient had no visual symptoms. The best-corrected visual acuity was 20/20 in both eyes. Ocular motility, anterior segment, and intraocular pressure were normal; no relative afferent pupillary defect was found. A translucent retinal lesion with a 2x3 papillary diameter, surrounded by areas of hyperpigmentation and atrophy of retinal pigment epithelium, was observed in the inferotemporal quadrant of the left eye. Diffuse vitreous deposits were present (Fig. 1, A and B); an atrophic lesion was present in the right eye in the superotemporal quadrant (1x1 papillary diameter). Electroretinogram was normal in both eyes.

On fluorescein angiography, the left eye showed an inferotemporal, well-demarcated, oval atrophic area of pigment epithelium; a similar lesion was present in the right eye (Fig. 1, C and D).

The general physical examination was negative.

Twelve years later, there were no detectable changes in the retinal lesions or in vitreous deposits.

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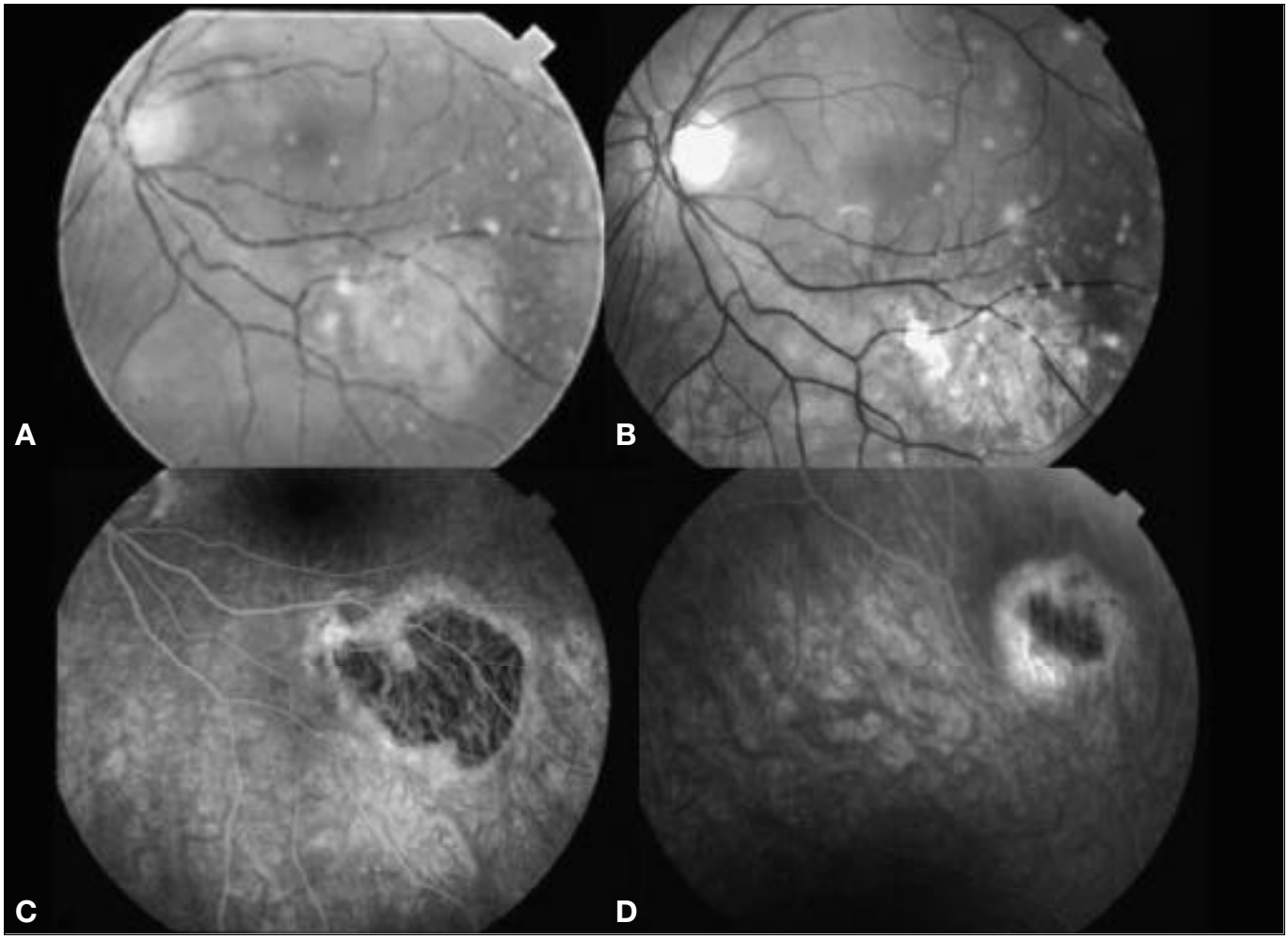

Fig. 1 - (**A–C**) *Left eye: clinical photograph shows retinoma associated with calcified vitreous deposits. Early phase fluorescein angiography shows an oval-shaped atrophic area of pigment epithelium, which allows for the visualization of choroidal circulation, while late-phase fluores cein angiography shows hyperfluorescent borders of the atrophic area.* (**D**) *Right eye: similar smaller lesion.*

DISCUSSION

Retinocytoma, first described by Gallie et al (4) in 1982, is a benign tumor that presents peculiar characteristic ophthalmoscopic features consisting of a translucent, gray lesion frequently associated with calcified foci, pigment epithelial hyperplasia, and irregular looping blood vessels. Some authors have proposed the term retinoma or retinoblastoma group O to define this tumor (5, 6).

Retinocytoma is estimated to occur in 2% to 10% of patients with retinoblastoma and their families. Although it usually does not grow, it may, nevertheless, undergo malignant transformation to retinoblastoma (7-9). The presence of vitreous seeds alone does not signify malignant transformation, which requires additional findings such as size and color changes of the tumor and appearance of fine vascularization on the surface of the mass. Furthermore, the vitreous seeding overlying a retinocytoma is calcified and bright while the vitreous seeding of an active retinoblastoma has a soft, cotton wool appearance.

Here we report a 35-year-old asymptomatic father of a child with bilateral retinoblastoma who presented a bilateral retinocytoma associated with calcified vitreous deposits in the left eye. Ophthalmoscopic findings in both eyes remained unmodified over 12 years of follow-up.

Similar cases have been reported by Lueder et al (1); they documented the presence of “focal vitreous opacities” in two adults with retinocytoma. The follow-up of these two cases has been 16 and 33 years and no change in the vitreous opacities has been noticed.

In a large series of patients with retinocytomas (17 cases) reviewed by Singh et al (2), localized vitreous deposits of calcium were observed in 7 (29%) of the tumors.

The mechanism of tumor regression in retinocytoma is unknown but might involve apoptosis (10). Regression caused by ischemic or immune-mediated necrosis is incompatible with the histopathologic features of retinoblastoma (11).

Tumor regression equally extends to the subretinal and vitreous seeds; we could speculate that an unknown factor could diffuse though the tumor components and induce a complete involution (12).

As reported in the literature (2, 4, 7, 9), any one of the three ophthalmoscopic characteristics of retinocytoma, such as the presence of a translucent retinal mass, calcification, and retinal pigment epithelial alteration, was documented in all retinocytomas.

Calcified vitreous deposits are a recently described feature of retinocytoma, which should be added to the triad of reported features.

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