Idiopathic Juxtafoveolar Retinal Telangiectasis and Pigment Epithelial Hyperplasia: An Optical Coherence Tomographic Study

One patient with bilateral idiopathic juxtafoveolar retinal telangiectasis (IJRT) revealing pigment hyperplasia in 1 eye was investigated by means of fluorescein angiography and optical coherent tomography (OCT).

Report of a Case. A 63-year-old man was referred to our department for gradual loss of central vision in both eyes. The visual acuity was 0.6 in right eye and 0.3 in the left eye. The Amsler test result was negative in both eyes. Biomicroscopic evaluation revealed in the right eye a small black stellate plaque temporal to the macula (Figure 1). In the left eye, a small intraretinal hemorrhage was revealed temporal to the macula. Early phases of fluorescein angiography showed minimal evidence of capillary dilation, while in the late phases, a mild staining surrounding the foveolar retina temporally was evident. This aspect was more marked in the right eye. The retina in the macular area of both eyes appeared flat by the OCT. The small plaque of pigment epithelial hyperplasia, in the right eye, was revealed by the OCT line scans as a highly reflective white-red area within the inner retina (Figure 2). A dark intraretinal area was detected under the hyperreflective band, due to its full screening effect, which produces a sort of barrier effect.

Comment. Based on different biomicroscopic and fluorangiographic patterns, Gass and Blodi1 have classified IJRT into 3 groups. Group 1 patients have occult telangiectasis and minimal exudation, and group 3 patients have easily visible telangiectasis and parafoveal capillary occlusion. Group 2 is often associated with superficial retinal crystalline deposits, right angle venules, and, sometimes, small plaques of pigment hyperplasia.

Retinal pigment proliferation and migration into the inner retina to form black stellate plaques was hypothesized by Gass and Blodi. However, a histological study showing the epithelial proliferation into the inner retina in some cases of retinal telangiectasis was never reported.

In this case of type 2 IJRT, the OCT line scans of the small plaque lesion showing the hyperreflective band (white-red pseudocolor)
within the inner retina could be consistent with the hypothesis reported by Gass and Blodi.

The potential of OCT examination for detecting macular diseases was already pointed out in other studies. The images reported herein confirm that OCT is a new modality that can be used in conjunction with other diagnostic procedures to study patients affected by macular diseases. Moreover, OCT images allow proper interpretation to specific morphologic alterations, as in the present case, where histological examination was not available.

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Diffuse and Circumscribed Choroidal Hemangiomas in a Patient With Sturge-Weber Syndrome

The Sturge-Weber syndrome is a rare, sporadic neuro-oculocutaneous disorder, the clinical features of which may include facial nevus flammeus (port-wine staining), buphthalmos, epilepsy, mental retardation, and hemiplegia. The most frequent ocular manifestation of the syndrome is a diffuse choroidal hemangioma, in contrast to the well-circumscribed choroidal hemangiomas seen in patients without the syndrome. To our knowledge, we report the first case of a diffuse choroidal hemangioma in one eye and a circumscribed choroidal hemangioma in the fellow eye of a patient with Sturge-Weber syndrome.

Report of a Case. A 3-year 8-month-old boy was first seen with a history of bilateral nevus flammeus involving the trigeminal regions since birth and was referred to our medical institution with a diagnosis of unilateral retinoblastoma. The child was noted by his parents to have an “in-turning right eye” since 2 months of age. There was neither history of eye pain nor photophobia. Visual acuity was 20/200 OD and 20/30 OS. A 4+ right afferent pupillary defect and a 60–prism diopter right exotropia were present. Slitlamp examination results were remarkable only for port-wine staining of the upper and lower eyelids bilaterally and leukocoria of the right pupil. Corneal diameters were within normal limits and there was no clinical evidence of buphthalmos. Findings from dilated funduscopic examination of the right eye revealed a diffuse choroidal hemangioma (with the classic “tomato ketchup” appearance), an overlying exudative total retinal detachment (Figure 1), retinal pigment epithelial mottling, and subretinal fibrosis. Results of dilated funduscopic examination of the left eye revealed a circumscribed choroidal hemangioma nasal to the optic disc without evidence of retinal complications (Figure 2). Examination under anesthesia showed intraocular pressures of 44 mm Hg OD and 18 mm Hg OS. The high intraocular pressure in the right eye was considered to be secondary to the diffuse choroidal hemangioma and resultant increased episcleral venous pressure. Echography revealed a diffuse...
choroidal lesion with high internal reflectivity and a total, open funnel-shaped retinal detachment in the right eye (Figure 3), and a circumscribed nasal choroidal lesion with high internal reflectivity in the left eye. Brain magnetic resonance imaging revealed a type I Arnold-Chiari malformation. Due to the long-standing detachment, the hemangioma in the right eye was not considered for treatment with external beam radiation. Serial follow-up evaluations (including examinations under anesthesia and a repeated brain magnetic resonance imaging) demonstrated no clinical, echographic, or neuroimaging changes throughout the 1-year follow-up interval.

Comment. Bilateral nevus flammeus, intracranial calcifications, and diffuse choroidal hemangiomas have been reported previously in patients with the Sturge-Weber syndrome,1-3 and there is one report of bilateral circumscribed choroidal hemangioma in an otherwise healthy woman.4 To our knowledge, we report the first patient with Sturge-Weber syndrome with a diffuse choroidal hemangioma in one eye and a circumscribed choroidal hemangioma in his fellow eye. The Sturge-Weber syndrome is classically associated with diffuse choroidal hemangiomas, the management of which is generally limited to treatment of associated complications such as glaucoma and retinal detachment. Our case demonstrates that patients with Sturge-Weber syndrome may also harbor circumscribed choroidal hemangiomas that warrant careful follow-up in order to detect promptly the need for treatment.

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