Discussion

The PCV lesion consists of a branching network of vessels in the inner choroid and aneurysmal vascular dilations at the border of the network of the vessels. There appear to be two types of PCV: an idiopathic entity occurring in younger people and a separate subgroup of patients in an older age group with these clinical symptoms, which may be confused with occult choroidal neovascularization (CNV). No underlying systemic or ophthalmic factor has been confirmed in its pathogenesis.[4] PCV has been described in association with hypertension and sickle cell retinopathy, and it has been postulated that the lesions may represent an insult to the choroidal vasculature.

Circumscribed choroidal hemangiomas are considered congenital, vascular, relatively rare hamartomas whose growth is related to the development of the choroid. These tumors generally are discovered in adulthood, when they become symptomatic because of the appearance of a serous retinal detachment and degenerative macular changes. ICG angiography can be helpful in confirming the diagnosis and in the clinical evaluation of the hemangiomas.[5]

We described a patient with concomitant PCV and circumscribed choroidal hemangioma and found some similarities between the two diseases. First, both of the lesions are related to the development of the choroidal and are seen as choroidal aneurysmal dilations. Second, they both are seen as a reddish orange, elevated lesion in the fundus. Third, they share some similar features during the course of fluorescein and ICG angiography. In addition, histologically, circumscribed choroidal hemangiomas consist of a mixture of small (capillary) or large (cavernous) vascular channels lined by flat endothelial cells separated by connective tissue septae.[6] Similar histologic characteristics are shared by PCV, which consists largely of vascular channels, fibrin around endothelium, and the elevated polypoidal and tubular choroidal lesions correspond to large, thin-walled, cavernous vascular channels.[6,7] Although there may be some analogous findings on ICG angiography and origination, there are more differences between the two entities. Specifically, choroidal hemangiomas are thought to be congenital lesions that may manifest later in life. Furthermore, they tend to be relatively stationary, although they may occasionally grow and exhibit increased exudation. They are thought to be true hematomas of the choroid.[5] Conversely, PCV likely represents a more reactive response of the choroidal vasculature. These lesions often show growth, remodeling, and occasional conversion to more acute types of CNV and manifest with a different spectrum of clinical findings, including hemorrhage and exudative detachment of the retina.[1,3,4,6] This rare case shows many similarities and also provides some clues regarding the pathogenesis of PCV. Therefore, we hypothesize that some cases of PCV may be related pathophysiologically to congenital vasculopathy, just like circumscribed hemangioma. Whether the association between the two vascular lesions is coincidental or the pathogenesis of PCV is related to a congenital condition necessitates further investigation.

Key words: internal limiting membrane, macular hole surgery, triamcinolone acetonide.

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SUCCESSFUL TREATMENT OF SEVERE RETINAL VASCULAR ABNORMALITIES IN INCONTINENTIA PIGMENTI

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Incontinentia pigmenti (IP), also known as Bloch–Sulzberger syndrome, is a rare disorder. Probably 800 cases of IP have been reported. IP is a generalized

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abnormality of ectodermal and mesodermal structures and is well recognized in dermatology for its characteristic skin disturbances. The skin lesions appear soon after birth as a linear pattern of erythema with vesicles or bullae on the trunk and extremities. The diagnosis is confirmed by a skin biopsy. IP is a multiorgan disease with dermatologic, dental, neurologic, and ocular abnormalities. The disease is inherited as an X-linked dominant condition and is normally lethal in males because of severe disturbances in the hematopoietic and immunologic systems. IP was shown to be associated with mutations in the NEMO gene, which maps to Xq28. The most serious manifestation of IP is central nervous abnormalities, which occur in approximately 30% of these patients.

Ocular abnormalities, often asymmetric, occur in 25% to 77% of patients[1] and include strabismus, pigmentation of the conjunctiva, nystagmus, lesions of the posterior segment, cataract, and optic atrophy. The retinal findings range from peripheral avascularity to preretinal fibrovascular proliferation with vitreous hemorrhage and tractional retinal detachment. Tractional retinal detachment has a poor prognosis and is difficult to manage.[2]

The purpose of this report was to describe successful treatment of proliferative retinopathy in IP with indirect diode laser coagulation and additional pars plana vitrectomy in a female infant.

Case Report

The female patient was born at term after an uneventful pregnancy. Hyperpigmentation and vesicular lesions of the skin appeared 7 days after birth. In computed tomographic angiography, vascular cerebral lesions with closure of multiple small vessels were evident. Her pediatrician and dermatologist established the diagnosis of IP based on her characteristic skin lesions. She had a positive family history for IP, which affected her mother, maternal grandmother, and her two sisters.

Ophthalmoscopic examination at the age of 4 weeks showed a few areas (3 clock hours) of peripheral retinal avascularity in the right eye. The fundus findings of the left eye included dilatation and tortuosity of the retinal vessels. A frond of new vessels had formed near the area of peripheral avascularity with a gray ischemic appearance (Figure 1). These findings were similar to those of zone 1 stage 3 retinopathy of prematurity in accordance with the international classification of retinopathy of prematurity. Fluorescein angiography was not performed. The left eye was also noted to have neovascularization involving the optic disk. There was no evidence of retinal detachment. Because of the risk of progressive retinopathy and possible tractional retinal detachment, the patient was scheduled for laser treatment. Indirect diode laser treatment of the peripheral avascular retina was performed 3 days later on both eyes (Figure 2) (right eye, 336 spots; left eye 1,974 spots; 0.1 seconds; 300–500 mW). The right eye was treated because of severe alterations in the left eye and to avoid possible additional surgery for the right eye.

On follow-up, in the less affected right eye, the retina was attached and laser scars were visible in the periphery. In the left eye, the ridge disappeared and the tortuosity was diminished. The neovascularization on the nasal side of the optic disk remained.

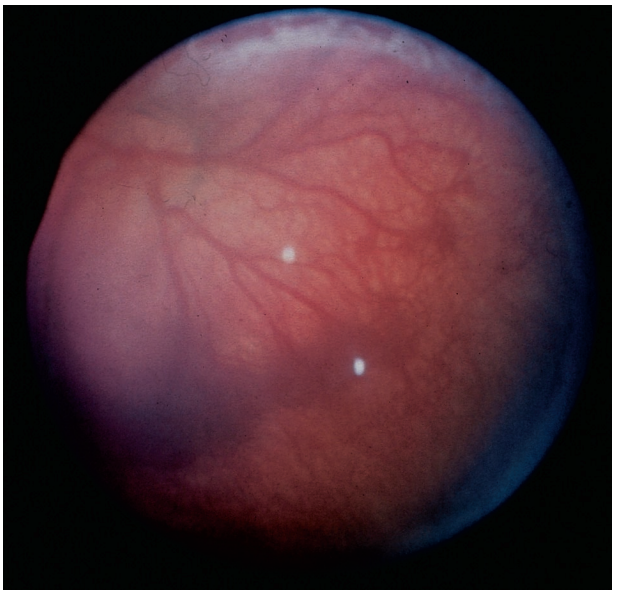

Fig. 1. Retinopathy in incontinentia pigmenti at the age of 4 weeks. From the disk to the near frond of vascularization, vessels are dilated. Proliferations are present at the border of the vascularized area in zone I.

Two months after laser treatment, a dense preretinal hemorrhage over the entire posterior pole occurred in the left eye with fibrovascular proliferations on the optic disk. No sign of regression occurred within 2 weeks. Because of the risk of amblyopia and a tractional retinal detachment, a pars plana vitrectomy was performed. The hemorrhage along with the peripheral vitreous traction, the central vitreous traction, and the fibrovascular traction on the optic disk were removed. No intraocular tamponade was necessary. At the 5.5-month follow-up, no additional bleeding or

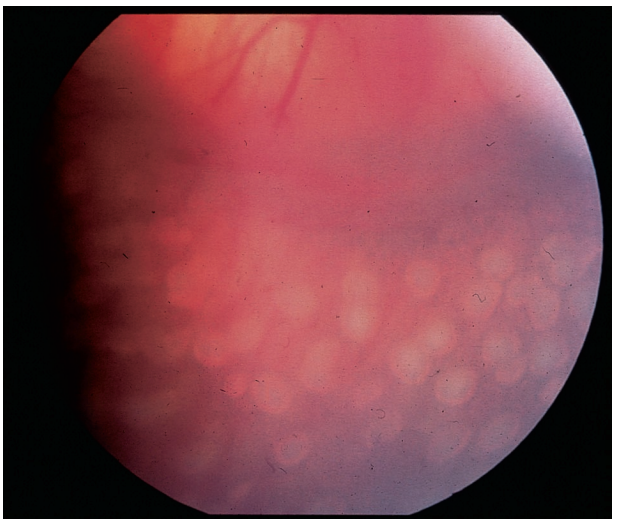

Fig. 2. The patient at the age of 4 weeks. Laser spots in the avascular area immediately after indirect laser treatment with a proliferative ridge between the vascular and avascular area.

fibrovascular traction had occurred. The retina remained attached with chorioretinal laser scars in the periphery. The anterior segment and lens appeared healthy. Cycloplegic refraction was +1.50 −0.50 100° in the right eye and −13.00 diopters in the left eye. Axial length was 22.2 mm in the right eye and 24.5 mm in the left eye. The patient developed a constant esotropia of the left eye measuring 30 prism diopters. She is suffering from seizures twice a week. Because of her mental retardation, only a visual acuity of fixation and following could be obtained. There was no afferent pupillary defect. Occlusion therapy of the right eye was tried for several months but was not tolerated.

Discussion

Ocular involvement in IP has been reported in 25%, 35%, and 77% of cases.[1] Severe visual impairment from retinal detachment was found in 39% of affected eyes in one series. In these eyes, ocular manifestations were detected too late for treatment.

The clinical course and fundus appearance are similar to those in retinopathy of prematurity. Analogous to the treatment rationale for retinopathy of prematurity, ablation of the peripheral avascular retina has been performed with variable success.[2–5]

An ophthalmologist should examine infants with a diagnosis of IP immediately, because severe retinal vascular abnormalities may be present already in the first month of life.

The indications and timing of intervention of the retinal changes in IP remain unclear, but coagulation treatment should be offered early for patients with large avascular areas and beginning fibrovascular proliferation in an attempt to stop progression toward a tractional retinal detachment. In eyes with additional vitreous hemorrhage resulting from fibrotic vascular remnants, a pars plana vitrectomy should be considered for therapy.

Key words: internal limiting membrane, macular hole surgery, triamcinolone acetonide.

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RETINAL PIGMENT EPITHELIAL TEAR IN A PATIENT WITH CENTRAL SEROUS CHORIORETINOPATHY TREATED WITH CORTICOSTEROIDS

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A patient with central serous chorioretinopathy who had a retinal pigment epithelial tear after inappropriate treatment with oral and subconjunctival steroids is described. It is known that corticosteroid therapy is associated with the development and exacerbation of central serous chorioretinopathy (CSC).[1,2] We report a case of CSC where rapid exacerbation of serous retinal detachment complicated by the development of a retinal pigment epithelium (RPE) tear after corticosteroid treatment occurred, thus providing further inferential evidence that corticosteroid therapy has an adverse effect on the severity and course of CSC.

Case Report

A 45-year-old man presented to an ophthalmologist because of a 2-week history of blurred vision in the right eye. His medical history was unremarkable. Results of examination showed that his best-corrected visual acuity was 20/28 in the right eye and 20/20 in the left eye. Fundus examination revealed an oval retinal elevation of 2.5 disk diameters at the posterior retina in the right eye (Fig. 1A) and subtle RPE discoloration inferonasal to the fovea in the left eye. Fluorescein angiography (FA) showed sharply demarcated hyperfluorescence that corresponded to retinal elevation and other small round-shaped hyperfluorescent spots without active dye leakage in the right eye (Fig. 1B) and a hyperfluorescent spot without active leakage that corresponded to RPE discoloration in the left eye (Fig. 1C). The ophthalmologist suspected bilateral choroiditis and prescribed 3 mg of oral dexamethasone daily for 1 week, which was decreased to 2 mg daily thereafter. The patient had also been treated with 2 mg of subconjunctival dexamethasone twice a week in his right eye. Two weeks later, visual acuity in the right eye deteriorated to 20/100, and serous retinal detachment was found in the right eye. He was referred to our hospital the next day.

At the time of examination, his best-corrected visual acuity was 20/200 in the right eye and 20/20 in the left eye. Fundus exami-

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