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MAJOR REVIEW

Purtscher's and Purtscher-like Retinopathies: A Review

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Abstract. Purtscher's retinopathy is a rare condition, seen in patients with a history of trauma or other systemic disease. The clinical appearance would suggest embolic occlusion of the pre-capillary arterioles as the most likely pathogenesis, although other factors leading to blood-vessel damage may also contribute. The diagnosis is made on clinical grounds and supported by intravenous fluorescein angiography. Without treatment, the majority of patients recover some of their visual function. Treatment with systemic steroids may improve visual outcome in some patients but at present there is little evidence to support such treatment routinely. (**Surv Ophthalmol 51**:129–136, 2006. © 2006 Elsevier Inc. All rights reserved.)

Key words. acute pancreatitis • complement activation • embolic vascular occlusion • Purtscher retinopathy

I. Introduction

Purtscher's retinopathy was first described in 1910 by Otmar Purtscher in a middle-aged man who fell from a tree onto his head and suffered a brief loss of consciousness.[37] Purtscher noted multiple areas of retinal whitening and hemorrhage in the posterior pole of both eyes. Despite initial visual loss, the acuity recovered without treatment. Since then, in addition to various types of trauma, a similar retinal appearance has also been described in a variety of conditions including acute pancreatitis, fat embolism syndrome, renal failure, childbirth, and connective tissue disorders (Table 1).[2,3,5,7–10,14,15,17,21,23–25,28,30,31,35,36,42,44–46,48] The term "Purtscher-like retinopathy" is sometimes used to describe the retinopathy seen in conditions other than trauma. However, in this article we have used the term *Purtscher's retinopathy* to describe the retinal appearance in all the above-mentioned conditions. Although the exact mechanism of injury remains unclear, current evidence suggests that it is embolic in nature.

II. Features

Patients with Purtscher's retinopathy present with loss of acuity in one or both eyes, ranging from minimal impairment to hand movements visual acuity. When a reliable history can be obtained, the visual disturbance may be delayed by 24–48 hours from the onset of the associated illness. Loss of acuity may be accompanied by field loss in the form of central, paracentral, or arcuate scotomata. Peripheral visual function is usually preserved.

Acute fundus abnormalities in Purtscher's retinopathy include *Purtscher flecken*, cotton-wool spots, retinal hemorrhage, and optic disk swelling. The

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TABLE 1

Purtscher's Retinopathy : Known Associations

Head Trauma
Long bone fracture, orthopedic surgery
Acute pancreatitis
Chest compression
Chronic renal failure
Hemolytic uremic syndrome
Childbirth
Connective tissue disorders, e.g., systemic lupus erythematosus
Cryoglobulinemia
Weight-lifting
Battered baby syndrome
Orbital injection

characteristic Purtscher flecken consists of multiple, discrete areas of retinal whitening in the superficial aspect of the inner retina, between the arterioles and venules. The flecken are polygonal and of variable size, ranging from a quarter to several disk areas (Figs. 1A and B). The retinal whitening may extend to the edge of an adjacent venule but a clear zone usually exists between the affected retina and an adjacent arteriole. A pseudo-cherry red spot may be seen when the retinal whitening surrounds the fovea. Retinal hemorrhages are often minimal and are typically flame-shaped but dot and blot hemorrhage may occur. These fundal abnormalities are confined to the posterior pole, namely within the macula and immediately nasal to the optic disc. Most cases are bilateral, although unilateral cases have been reported.[9,28,31,48] The acute changes in the retina may not be apparent until 24–48 hours after the systemic illness. Without treatment, they resolve spontaneously within 1–3 months, and may be replaced by mottling of the retinal pigment epithelium, temporal disc pallor, and attenuation or sheathing of the retinal vessels (Figs. 1E and F).

On intravenous fluorescein angiography (IVFA), the choroidal fluorescence may be masked by the retinal whitening or blood, non-perfusion of the smaller retinal arterioles or capillaries may be seen and there may be late leakage from the retinal vessels in areas of ischemia (Figs. 1C and D).[3,24] Leakage from the optic nerve has also been reported.[46] Areas of choroidal hypofluorescence have been noted on indocyanine green angiography, implying the additional involvement of the choroidal circulation.[15] Further evidence of choroidal involvement in Purtscher's retinopathy comes from histopathology, electroretinography, and the late changes at the level of the RPE described previously.[17,26,46]

In most cases, the fundus appearance and an associated systemic illness will be sufficient to make the diagnosis. The differential diagnosis includes branch or central retinal artery occlusion, commotio retinae, and fat embolism. However, in Purtscher's retinopathy, the symptoms and signs are often delayed relative to the systemic event, there is no direct globe trauma, visible retinal emboli are absent, and the retinal whitening spares the retina immediately adjacent to the retinal arterioles.

III. Pathogenesis

In his original report, Purtscher postulated that the retinal changes were due to an extravasation of lymph from retinal vessels following the sudden increase in intracranial pressure secondary to head injury. Since then a variety of alternative mechanisms have been suggested in the pathogenesis of the Purtscher flecken and other retinal lesions (Table 2). Impaired venous return and acute expansion of retinal veins secondary to chest compression, weight-lifting, battered baby syndrome, and asphyxia may produce retinal hemorrhage, cotton-wool spots and, occasionally, lesions resembling Purtscher flecken.[25,32] Alternatively, a retinal vasculitis may be induced by lipase after systemic injury and lead to thrombosis and vascular occlusion.[11,38] Although the exact mechanism remains unknown, the multifocal lesions restricted to the posterior pole, bilateral involvement and occlusion of the retinal vessels on fluorescein angiography all suggest that the pathogenesis involves embolic occlusion of the precapillary arterioles as the most likely cause of the flecken.

The characteristic Purtscher flecken occur in the inner retina, between the retinal arterioles and venules. The clear zone apparent between the affected retina and an adjacent arteriole corresponds to the capillary free area, extending for an average of 50μ on either side of the retinal arteries and precapillary arterioles.[34] The presence of this clear zone would suggest that the primary pathology lies within the precapillary arterioles. Proximal occlusion of the retinal arteries by large emboli would be expected to produce the confluent retinal whitening seen, for example, in branch arterial occlusion, whereas distal occlusion of the retinal capillaries alone by small emboli would lead to the development of a cotton-wool spot. Varied lesions have been induced experimentally in cats and pigs by the injection of glass spheres and also by fibrin clots, and platelet and leukocyte aggregates of varying size.[1,4,12,29,40,41] Intermediate-sized emboli were required to produce lesions resembling the typical Purtscher flecken.

Potential emboli in the pathogenesis of Purtscher retinopathy include air, fat, leukocyte aggregates,