24 Vasculopathies with Acute Systemic Diseases

24.1 Purtscher’s Retinopathy

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Core Messages
- Purtscher’s retinopathy is a traumatic retinal angiopathy and was originally described in individuals with severe head trauma.
- Symptoms include rapid and painless vision loss.
- Usually occurs in both eyes, but unilateral cases have been reported.
- Acute funduscopic findings include cotton-wool spots and intraretinal hemorrhages primarily located in the posterior pole. Optic disk swelling may also occur.
- Purtscher’s-like retinopathy can occur in other traumatic conditions (compressive chest injuries, long bone fractures), collagen vascular diseases (systemic lupus erythematosus, scleroderma, dermatomyositis), kidney diseases, and childbirth.
- Treatment is supportive and directed at the underlying medical condition.

24.1.1 History

Purtscher’s retinopathy was first described by Otmar Purtscher in 1910 as a syndrome of multiple, white retinal patches, superficial retinal hemorrhages, and papillitis occurring in five patients with severe head trauma [11]. He originally named this condition *angiopathia retinæ traumatica* and hypothesized that white, superficial retinal patches were lymphatic extravasations caused by a sudden increase in intracranial pressure secondary to massive head trauma.

The term Purtscher’s retinopathy or Purtscher’s-like retinopathy is currently used to describe similar fundus findings associated with various conditions: trauma (head trauma, compressive chest injuries, long bone fractures with fat embolism), collagen vascular diseases (systemic lupus erythematosus, scleroderma, dermatomyositis), kidney disease, and childbirth (amniotic fluid embolism) [2, 4, 5, 8, 12].

Although associated with several systemic conditions, Purtscher’s retinopathy is a relatively uncommon finding and published reports in the literature have been limited to case reports.

24.1.2 Special Pathologic Features

Essentials
- Complement-induced retinal leukoembolization is likely responsible for the ophthalmoscopic findings in Purtscher’s retinopathy.

Although Otmar Purtscher initially postulated that the white, superficial retinal patches (now recognized as cotton-wool spots) were lymphatic extravasations caused by the sudden increase in intracranial pressure seen in severe head trauma [11], the exact pathogenesis of Purtscher’s retinopathy remains unknown. Several authors have hypothesized that microembolic events, such as fat embolization in long bone fractures and air embolization from compressive chest injuries, may be responsible [3]. Other scientists speculate that venous reflux with capillary engorgement of the upper body plays a role [10].

More recently, studies have shown that leukocyte aggregation by activated complement factor 5 (C5a) may lead to leukocyte emboli that can occlude the peripapillary retinal capillaries in conditions such as trauma, acute pancreatitis, and collagen vascular diseases [7]. Several authors have postulated that complement-induced retinal leukoembolization occurs in the setting of Purtscher’s retinopathy, and animal models have shown evidence of multiple small retinal arteriolar occlusions [6, 9].
24.1.3 Clinical Course of the Disease

**Essentials**
- Painless, rapid vision loss
- Frequently bilateral involvement
- Acute funduscopic findings include cotton-wool spots and intraretinal hemorrhages located primarily in the posterior pole
- Fluorescein angiography may demonstrate retinal arteriolar occlusions, capillary non-perfusion, and leakage from vessels in the affected area

Purtscher’s retinopathy is most often bilateral, but it may be reported to occur in unilateral cases [3]. It may occur after traumatic injuries or a Purtscher-like retinopathy may be associated with other systemic conditions such as collagen vascular disease, kidney disease, and childbirth [2, 4, 5, 8, 12].

Acute funduscopic findings include cotton-wool spots and intraretinal hemorrhages, primarily located in the posterior pole (Fig. 24.1.1). Optic disk swelling may be present. Fluorescein angiography may reveal focal retinal arteriolar occlusions in the perifoveal area, capillary non-perfusion, and leakage from vessels in the areas of infarction (Fig. 24.1.2) [5].

Visual acuity is markedly decreased in the affected eye(s) and may range from 20/200 to light perception. An afferent papillary defect may be present if the condition is unilateral or asymmetric.

24.1.4 Differential Diagnosis

The differential diagnosis of Purtscher’s retinopathy includes retinal vascular disorders that can result in intraretinal hemorrhages and cotton-wool spots. Central retinal vein occlusion, hypertensive retinopathy, diabetic retinopathy, Valsalva retinopathy, and lupus retinopathy may all present with intraretinal hemorrhages and nerve fiber layer infarcts.

Differentiating between Purtscher’s retinopathy and other conditions requires a thorough medical history to document preexisting medical conditions, such as diabetes and hypertension, and possible precipitating events, such as trauma, that may suggest one disease over the other. In addition, funduscopic findings such as lipid exudates, tortuous and dilated veins, and retinal neovascularization are not typical...