Purtscher’s Retinopathy

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Case 1
A young male presented with loss of vision in left eye following a road traffic accident. There was history of a heavy object falling on the chest at the time of the accident. He did not sustain injury to other organs or bones, but was unconscious for few hours and was kept under observation in the hospital for few days.

Vision at presentation was 6/6 in right eye and CFCF in left eye. Anterior segment examination was unremarkable.

Left eye fundus showed patches of opaque retina in the posterior pole with relative clearing around the vessels. There were few areas of preretinal hemorrhage also (Fig-1). Right eye fundus was normal (Fig-2).

Case 2
A female presented with decreased vision in both the eyes eighteen days after a normal vaginal delivery. The vision started deteriorating two days postpartum and gradually worsened to become CFCF both eyes. There was history of Pregnancy induced hypertension and post partum psychosis. Fundus examination showed patches of pale retina with pre-retinal hemorrhage in left eye and opaque retina in posterior pole resembling CRAO in right eye (Fig-3 & 4). Detailed cardiovascular evaluation and coagulation profile were normal.

Discussion
The clinical presentations in these patients describe a rare ischemic retinopathy called Purtscher’s retinopathy. The first patient was initially diagnosed as combined CRAO and CRVO and the second patient was referred to us with a diagnosis of CRAO.

Both the patients received treatment with high dose IV methylprednisolone, with no benefit.

Purtscher’s retinopathy
Purtscher’s retinopathy was named after an Austrian ophthalmologist, Othmar Purtscher.\[1,2\] He described this in 1912, as a cause of sudden vision loss in patients with head trauma, characterized by multiple white retinal patches and retinal hemorrhages.

Since this first report, similar retinal appearance has been observed in many nontraumatic conditions also. The associations of Purtscher’s retinopathy reported are:
• Head trauma. [2]
• Long bone fracture. [4, 5]
• Compressive Injuries of trunk & Barotrauma [4]
• Acute pancreatitis. [6]
• Child birth. [8]
• Renal Failure. [9]
• Steroid injections in and around the eye & nasal passages. [10]
• Retrobulbar anesthesia. [11]
• Connective tissue and vasculitic diseases. [12]
• SLE, TTP
• HELLP syndrome
• Amniotic fluid embolism.
• Purtscher’s retinopathy is usually bilateral, but can also be unilateral. Decreased vision occurs in the affected eyes, generally in the range of 20/200 to counting fingers. Vision may improve gradually depending on the severity of the retinal changes. Purtscher’s retinopathy associated with pre eclampsia has demonstrated a poor visual outcome. [8]

Pathophysiology
The exact pathogenesis of Purtscher’s retinopathy still remains controversial and various hypotheses have been put forward.

1. Purtscher described the white retinal patches (Purtscher’s flecken) as intraretinal changes resulting from extravasation of lymph from the retinal vessels due to sudden increase in intracranial pressure during severe head injury.

2. The most accepted mechanism is the complement activation induced leukoembolization. Activation of complement C5a following trauma, acute pancreatitis and vasculitic diseases, leads to aggregation of granulocytes leading to arterial occlusion and infarction of the microvascular bed.

3. Fat embolism in cases of long bone fractures and pancreatitis from enzymatic digestion of omental fat, amniotic fluid embolism at the time of child birth or postpartum also leads to infarction of the retina.

4. Other mechanism is angiospasm, caused by acute rise in venous pressure from compressive chest injuries and head injuries, resulting in endothelial cell damage due to sudden rise in intraluminal pressure.

Clinical features
The patients present with sudden or gradual, unilateral or bilateral loss of vision. Fundus examination shows cotton-wool spots due to obstruction in the axoplasmic flow. Large areas of retinal whitening, having typical polygonal shape with small strips of clearing adjacent to large retinal blood vessels, termed Purtscher’s flecken are characteristic. Intraretinal or pre retinal hemorrhages are also noticed. Rarely optic disc edema can occur. After 4 to 6 weeks the white retinal patches fade away and eventually the fundus may appear normal, though there may be residual pigmented mottling and optic atrophy.

Fluorescein Angiography is often not indicated. It is reported to show features like capillary non-perfusion, late leakage from injured retinal vessels, retinal or disc edema and perivascular staining.

Treatment
Hyperbaric oxygen, intravenous methyl prednisolone, sub tenon Triamcinolone etc have been tried as treatment options. [13, 14, 15] Investigating for the underlying cause and treatment of the same whenever possible should be done wherever indicated. No treatment has been proven to be superior to observation for Purtscher’s retinopathy. Spontaneous visual recovery of at least two Snellen lines has been reported in 50% of the cases. There is some evidence that poor initial and final acuities may be associated with persistent acute changes in the retina. [3]

References