

CLINICAL IMAGES

Sudden loss of vision: Purtscher retinopathy in multiple myeloma

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A 53-year-old man presented with acute, severe loss of vision in both eyes. Three days earlier, he had noticed a painless deterioration of his vision, which had progressed until he could no longer read a newspaper using either eye. On examination, the patient's best corrected visual acuity was 20/100 using both eyes. He had full visual fields by confrontation and the motility of extraocular muscles was normal. No afferent pupillary defect was evident. Examination of the posterior segment showed bilateral edema in the optic nerve head with multiple cotton wool spots and areas of retinal whitening (Purtscher flecken) (Figure 1). Intravenous fluorescein angiography showed severe ischemia of the nasal macular retinal arterioles with extensive secondary late leakage. Based on these findings, we diagnosed Purtscher disease or Purtscher-like angiopathic retinopathy.

Pertinent laboratory findings included an elevated erythrocyte sedimentation rate of 83 (normal 0–20) mm/hr, elevated total protein of 92 (normal 60–80) g/L and a serum IgG immunoglobulin of 41.8 (normal 7–17) g/L. These results prompted a skeletal survey that showed small lytic lesions in the proximal humerus, skull and proximal femur. Serum protein electrophoresis showed a monoclonal M spike of 3.39 g, which was eventually determined by immunofixation to be IgG/Kappa type. His peak serum viscosity was elevated at 2.3 (normal 1.4–1.8) centipoises.

A bone marrow biopsy was then performed and showed large aggregates of plasma cells comprising approximately 33% of the cell mass. Immunohistochemical stains showed that the plasma cells were kappa light-chain restricted, which was consistent with multiple myeloma, and involved about 70% of the marrow cellularity.

Our patient presented with the classic appearance of Purtscher-like retinopathy as an initial manifestation of multiple myeloma. We considered the possibility that hyperviscosity from multiple myeloma was playing a role in our patient's

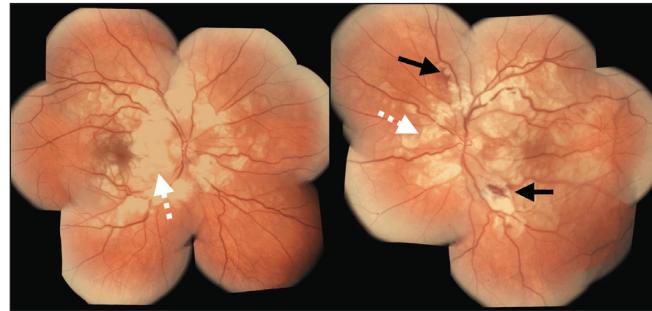


Figure 1: Composite digital fundus photographs of the posterior poles of the right and left retinas of a 53-year-old man with Purtscher retinopathy, showing areas of anterior retinal whitening (dotted arrows) with minimal flame hemorrhages (black arrows) and normal peripheral retina.

visual loss. Symptoms of hyperviscosity usually appear when a normal serum viscosity climbs to 4–5 centipoises, which often corresponds to a serum IgG level of 40 g/L.¹ The patient was treated with three cycles of plasmapheresis, resulting in a drop in the viscosity to a baseline of 1.3 centipoises. He was also given dexamethasone and lenalidomide. After eight months, his vision had improved to 20/400 (right eye) and 20/30 (left eye).

Purtscher's retinopathy is rare and most commonly seen after trauma, acute pancreatitis, crush-type injury, long-bone fracture, orthopedic surgery and childbirth.² Acute retinal signs often persist for at least one month after the initial injury and include Purtscher flecken, cotton wool spots and retinal hemorrhage.² Spontaneous visual recovery of at least two Snellen lines occurs in half of patients.²

This article has been peer reviewed.

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