HISTORY & EXAM:
A 53 year-old male violinist presented with a five day history of progressive, painless loss of vision in both eyes. He denied headaches, diplopia, transient visual obscurations or other neurological symptoms. He was vegetarian, in good health, and denied systemic disease.

A retina specialist thought he had bilateral papilledema and sent him for neuro-ophthalmology consultation. Examination showed that the best corrected visual acuity was 20/200 in the right eye and 20/40 in the left eye. The pupils were equal and reactive to light and there was a questionable right relative afferent pupillary defect. Confrontation visual field testing showed that he could count fingers in all quadrants of both eyes. Goldmann perimetry of the right eye, showed a moderately dense central scotoma, an enlarged blind spot, and slightly constricted peripheral isopters. In the left eye, Goldmann perimetry showed minimal constriction of the isopters superotemporally. With the Amsler grid, the patient noted distortion and waviness of lines superotemporally, O.S. The patient was able to identify 12 out of 15 Ishihara plates with the right eye, by fixating eccentrically with his nasal field. He identified all 15 color plates with his left eye. Extraocular movements were full and his slit-lamp examination was unremarkable. Intraocular pressures were within normal limits.

Examination of the ocular fundi revealed that both optic discs were edematous, elevated and surrounded by multiple flame-shaped hemorrhages that radiated from the optic discs and obliterated the disc margins. Small exudates and scattered dot and blot hemorrhages were present surrounding both optic discs; no substantial hemorrhage was noted along the retinal vascular arcades outside the immediate peripapillary area. In both eyes, the retinal venules appeared distended and engorged, with boxcarring of the blood column. There were serious macular detachments in both eyes. There were no abnormalities in the retinal periphery of either eye.

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:
Multiple Myeloma presenting with bilateral central retinal vein occlusions secondary to hyperviscosity syndrome

SUMMARY OF CASE INCLUDING PATHOLOGY:
The patient was admitted to the hospital for additional evaluation. Laboratory tests showed mild pancytopenia (WBC 3.1 K/microliter, Hgb 9.7 g/dL, platelets 135 K/microliter), high serum protein, low serum albumin, and high LDH. Vitamin B12, folate and iron levels were normal. CT of the head without contrast did not show any acute abnormality. MRI, MRA and MRV of the brain without contrast, revealed a poorly-defined heterogeneous infiltrate involving the clivus, suggesting a possible malignancy. A whole body bone scan was then obtained and showed multiple areas of uptake involving ribs, bilaterally, the right scapula, and left femur. A metastatic bone survey revealed vague lucencies throughout the skull consistent with multiple myeloma. Serum electrophoresis with immunofixation revealed a large IgA lambda monoclonal protein in the beta region. The patient’s serum viscosity was elevated at 7.24 centipoise (cps). A bone marrow biopsy established the diagnosis, showing diffuse infiltration by monoclonal plasmacytic cells precursors (CD38+, CD56+, and cytoplasmic lambda +). The patient underwent two courses of plasmapheresis in addition to treatment with lenalidomide, allopurinol, zoledronic acid and dexamethasone.

One month later, the visual acuity was 20/25 in both eyes, with significant absorption of the retinal hemorrhages and amelioration in the appearance of the retinal vasculature. Serous detachment in both maculae was still present. At six months, the patient’s visual acuity was 20/20 in both eyes with resolution of the retinal hemorrhages and normalization of the caliber of the retinal venules. There was no macular edema or serous detachment although there were residual hard exudates and minimal elevation of the optic discs, nasally. The patient has now undergone an autologous bone marrow transplant and is doing well.

- Bilateral Disc Swelling and visual loss was initially considered to be Papilledema.
- MRI showed infiltration of the clivus, suggesting a malignancy, possibly multiple myeloma.
- Further laboratory evaluation revealed hyperviscosity syndrome and a large IgA lambda monoclonal protein, confirming the diagnosis.
- The clinical findings of disc swelling, retinal hemorrhage and serous macular detachment were a result venous congestion resulting from the hyperviscosity syndrome caused by multiple myeloma.
- Only 2 such cases have been described previously in the literature (1, 2).

KEY WORDS: Bilateral Optic Nerve Swelling, Peripapillary Hemorrhage, Central Vein Occlusion, Hyperviscosity Syndrome, Multiple Myeloma

REFERENCES: