A 48-Year-Old Female with Progressive Visual Loss of the Left Eye

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Introduction:

A 48 year-old female referred for worsening left eye vision for several months and chronically poor vision in the right eye from glaucoma.

Exam:

Best corrected visual acuity was 20/50 in the right eye and 20/200 in the left eye. Intraocular pressures were within normal limits. No afferent pupillary defect was appreciated. Visual fields were full to confrontation and extraocular motility was full in both eyes. Anterior segment exam was notable for normal findings.

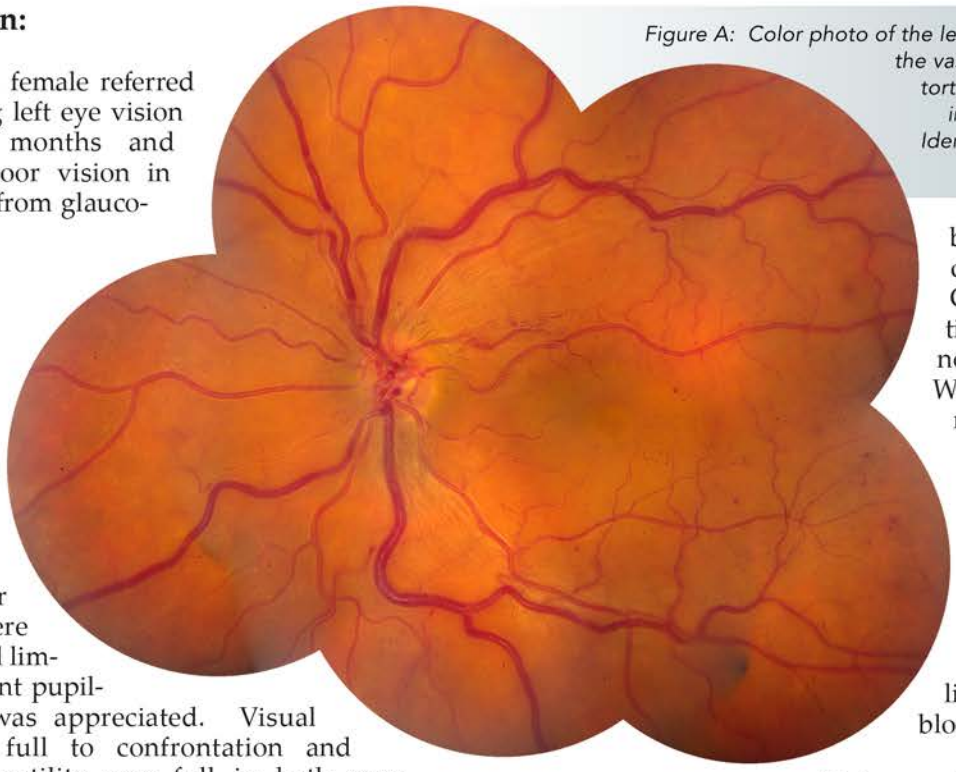

Figure A: Color photo of the left posterior pole. Notice the vascular engorgement and tortuosity along with diffuse intraretinal hemorrhages. Identical findings were seen in the left eye.

Dilated fundus examination revealed bilateral vascular engorgement and tortuosity along with diffuse intraretinal hemorrhages (Figure A). Macular examination was notable for a disciform macular detachment. OCT imaging confirmed this sub-macular fluid along with diffuse CME and exudate.

Discussion:

Our patient was sent to obtain urgent work up by her primary care physician. Her labwork revealed a blood protein concentration of 13.4 (normal 6.2 – 8.3) and globulin level of 8.2 (normal 2.2-3.9). A bone marrow biopsy confirmed 7% clonal CD45+ B-cell population and she was diagnosed with Waldenstrom macroglobulinemia (WM). R-CHOP (Rituximab, Cyclophosphamide, Oncovin, Prednisone) therapy was initiated with gradual decrease in her globulin levels and overall blood viscosity.

This case highlights the retinal changes seen in WM. This condition is a lymphoproliferative B-cell disorder that results in a hyperviscosity syndrome caused by an overproduction of monoclonal IgM antibodies.[1] WM has an annual incidence of 3.8 per 1 million people and accounts for approx. 2% of all hematologic malignancies.[2] The average age of onset is 63 years.[2] Seventeen percent of all patients with WM will show clinical symptoms as a result of hyperviscosity.[3] These symptoms can range from fatigue and weight loss, to neurological symptoms

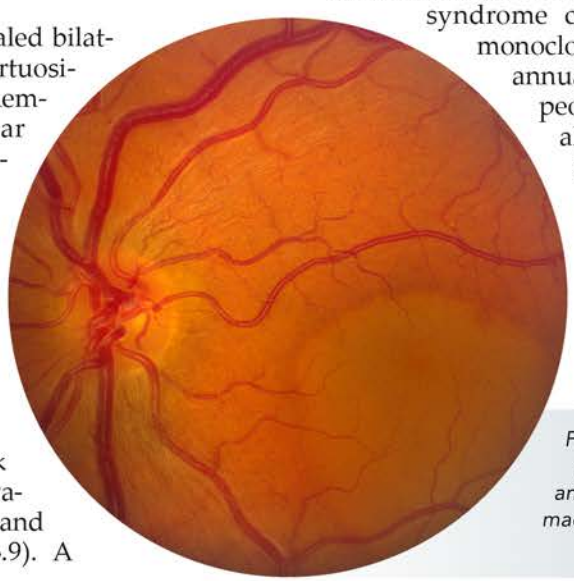

Figure B: Color photo of the left macula. Again, notice the vascular engorgement and tortuosity along with a large disciform macular detachment. The macular changes were only seen in the left eye.

and coma, and vascular disturbances such as spontaneous hemorrhages and retinopathy. The most common retinal findings include vascular dilation and central retinal hemorrhages. Several studies have concluded a direct correlation between retinal findings and the overall hyperviscosity state.[4,2] Higher immunoglobulin levels result in a larger mean venous and arterial diameters and a lower mean venous blood flow secondary to higher viscosity of the blood. The most common findings seen were peripheral and central dot-blot hemorrhages, dilated retinal vessels, and optic disc edema. Serous macular detachments are rarer and on average appear when mean immunoglobulin levels reach approx. 6497 mg/dl and a mean serum viscosity of 5.5 centipoise. Serous macular detachments are most likely form through a combination of venous stasis retinopathy and choroidopathy that eventually leads to a breakdown of the blood-retinal barrier and accumulation of IgM proteins in the sub-retinal space.[2] Treatment for this retinopathy focuses on systemic treatment of WM through the use of plasmapheresis, chemotherapy, blood transfusions, or transplantation of hematopoetic cells.[2] As overall IgM levels decrease, the hyperviscosity of the blood and venous stasis will improve. Macular CME or sub-retinal fluid should be observed and may partially or completely resolve as blood viscosity and flow normalize. Anti-VEGF or steroid therapy is not indicated as this is not a VEGF or inflammatory driven process. Unfortunately, resolution of retinopathy can leave behind vast areas of atrophy and subsequent poor visual acuity. The visual prognosis of this condition is guarded and highly influenced by the degree of retinopathy during the hyperviscosity phase.

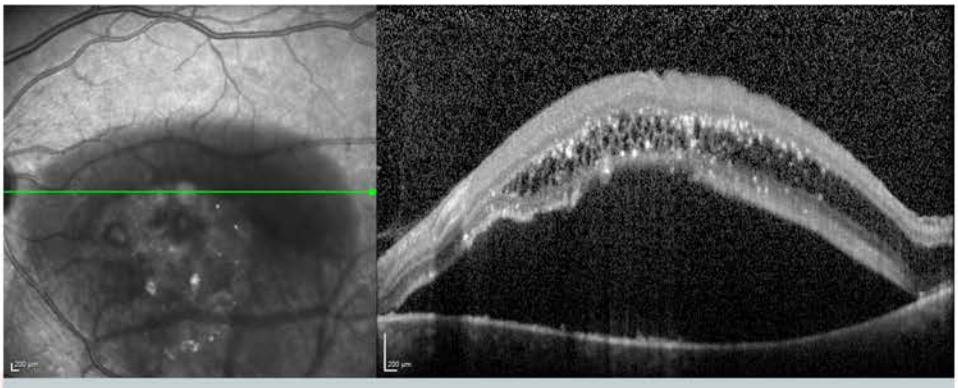

Figure C: OCT of the left macula. Notice the sub-macular fluid along with diffuse CME and exudate.

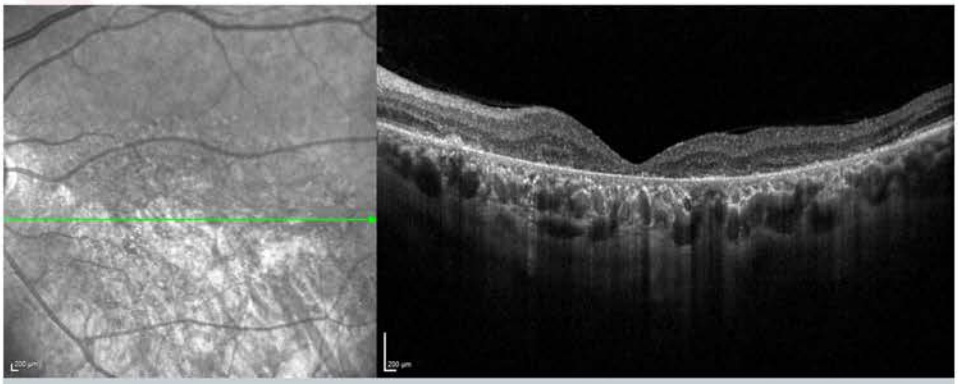

Figure D: OCT of the left macula eight years later. Notice the resolution of CME and sub-retinal fluid and presence of diffuse outer retinal atrophy.

References:

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