Abstract

Purpose: Wäldenstrom Macroglobulinemia (WM) is a non-Hodgkin lymphoma characterized by the proliferation of IgM-producing B cells which can cause hyperviscosity syndrome (HVS) and, ultimately, end-organ damage. Herein, we describe a case of a patient with long-standing WM who presented with signs of atypical central retinal vein occlusion (CRVO) and a salmon-patch conjunctival lesion that heralded relapse of his WM and concern for conversion to lymphocytic lymphoma.

Case: A 66-year-old male with history of controlled WM, presented with a unilateral decrease in vision and red eye. Anterior segment examination showed an elevated, pink-colored, conjunctival nodule in the right eye. Dilated fundus examination of the right eye revealed +2 optic disc edema, white-centered and flame-shaped retinal hemorrhages in the periphery, and macular edema. The atypical presentation associated with the salmon patch and pseudo-Roth spots were consistent with a diagnosis of WM Retinopathy. Under consultation with his oncologist, Bendamustine and Rituximab infusions were initiated. His ocular findings resolved within six weeks, but he succumbed to his underlying disease within six months of diagnosis.

Conclusion: Early recognition of worsening WM by ophthalmology allowed for appropriate and timely systemic restaging. Notably, there was rapid improvement in visual acuity, conjunctival lesion, and optic disc edema following the initiation of treatment in this case.