Session X: Disease Related Morbidities in

Waldenström's Macroglobulinemia

Abstract 157

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Hyperviscosity Related Retinopathy in Waldenström's Macroglobulinemia.
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Background: Waldenström's macroglobulinemia (WM) is characterized by an overproduction of immunoglobulin M (IgM), which can lead to a hyperviscosity syndrome (HVS) and HVS-related retinopathy. The earliest retinal changes due to HVS are peripheral hemorrhages and venous dilation. Retinal hemodynamic assessment has shown a good correlation between venous diameter and IgM level, and has documented the effectiveness of plasmapheresis for the treatment of HVS-related retinopathy. Recent study results are presented. Methods: 46 patients with WM were evaluated using indirect ophthalmoscopy with scleral depression, retinal photography, laser Doppler retinal blood flow measurements, and serum IgM and SV determinations. A retinopathy severity scale was developed, and the associated IgM and SV values were related to particular morphologic changes. In addition, nine patients with confirmed HVS due to WM were evaluated before and after plasmapheresis treatment. Results: Patients exhibited far-peripheral hemorrhages and venous dilation with increasing SV and IgM values. Central retinal changes were associated with significantly higher SV values. Retinal vessel diameter increased with increasing serum IgM and SV levels. The mean IgM level of patients with the earliest retinal changes was 5442 mg/dL. The mean SV level was 3.1 cp. Plasmapheresis resulted in significant reductions in serum IgM (46.5% ± 18.0%; $P = 0.0009$) and SV (44.7% ± 17.3%, $P = 0.002$). HVS-related retinopathy improved in all patients after plasmapheresis. After treatment, the venous diameter decreased in each patient by an average of 15.3% ± 5.8% ($P = 0.0001$). The percentage decreases in SV in the patients were significantly correlated with the percentage decreases in venous diameter ($P = 0.031$, $R^2 = 0.51$). Conclusion: Retinal manifestations of HVS occur at lower serum IgM and SV levels than previously reported. Indirect ophthalmoscopy with scleral depression along with retinal vessel diameter measurements are able to detect the earliest HVS–related retinal complications. Plasmapheresis is effective in reversing HVS-related retinopathy and in reducing abnormal venous dilatation.