

Retinal Manifestation in Multiple Myeloma

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Purpose: To show retinal findings in a patient with hyperviscosity syndrome secondary to multiple myeloma.

Case Report: A 60-year-old white woman had come to Hospital Alemão Oswaldo Cruz with seizures, mental confusion, weakness, epistaxis, hear loss and cyanosis. She was admitted to icu (intensive care unit) for hypovolaemia. The patient was initially treated with vasoactive drugs.

Pertinent laboratory findings included a elevated serum IgG immunoglobulin of 4385 mg/dL (normal 600-1500 mg/dL) and serum protein electrophoresis showed a monoclonal M spike of 5.24 g.

A bone marrow biopsy was then performed and showed large aggregates of plasma cells. Immunohistochemical stains showed that the plasma cells were kappa light-chain restricted (CD 138+ 10-20%, 90% Kappa positive), which was consistent with multiple myeloma. Bence-Jones proteins by immunofixation electrophoresis were positive.

A skeletal survey shows occult lytic lesions at T5, T6 and T9 vertebrae.

The hematology sector called for ophthalmologic evaluation thinking in hyperviscosity syndrome. Posterior segment examination showed numerous large retinal hemorrhages throughout the pole posterior and midperiphery along with mild vascular tortuosity in both eyes.

The patient was submitted to 5 five session of plasmapheresis at the beginning of treatment and after started chemotherapy schedule (Velcade, cyclophosphamide and dexamethasone). Until this moment, the patient visual acuity was 20/25 in both eyes after plasmapheresis with significant improvement of systemic findings.

Discussion: Multiple myeloma (MM) is a rare cancer characterized by the proliferation of malignant B cells (plasma cells) in the bone marrow and subsequent overabundance of antibodies.¹⁻³ The monoclonal protein is IgG in 50% of cases, IgA in 25% of cases, and IgD or IgE much less commonly. Immunoglobulin G (IgG) is usually the elevated protein in MM. ^{1, 2,3} Myeloma patients with retinopathy generally have significant reduced hemoglobin and platelet counts. The presence of retinopathy is not associated with a worse prognosis of MM and improves with systemic treatment. Common signs are discrete flame and Roth spot hemorrhages, cotton wool spots and microaneurisms. Hyperviscosity syndrome is rare in MM a can occur with signs similar to Waldenstrom macroglobulinemia. That signs included: dilated and tortuous retinal veins, superficial and deep retinal hemorrhages along with retinal edema. Central retinal and branch retinal vein occlusion can be associated with hyperviscosity retinopathy. ^{4,5}

Conclusion: hyperviscosity syndrome is a rare manifestation of multiple myeloma (MM) and when it occurs, can present concomitant retinopathy, with large hemorrhages and tortuous and dilated veins. There is association between central and branch vein occlusion with hyperviscosity syndrome. The retinopathy can improve with systemic treatment with plasmapheresis, and in the case of multiple myeloma, chemotherapy schedule.



Pictures: dilated funduscopy exam of both eyes at the unit care center showing numerous large hemorrhage throughout the pole posterior and midperiphery with mild vascular tortuosity and dilated veins.

References:

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