Copper Deposition in Cornea and the Lens Capsule as an Ocular Manifestation of Multiple Myeloma

Mina Naderi* and Mohsen Sfandbod
Department of Ophthalmology, Tehran University of Medical Sciences, Iran

Abstract
A 50 year-old woman presented to the hospital with a complaint of bilateral gradual progressive visual loss since three years ago, associated with cloudy discoloration of cornea in both eyes with copper deposition in the Descemet's membrane and deep layers of central cornea along with anterior lens capsule showed on Slit-lamp examination with other normal physical examinations. Through the pattern of copper deposition as previous reported other evaluations were around plasma cell disorders such as multiple myeloma. Laboratory evaluations demonstrated hypercupremia and increased ESR and mild anemia and increased monoclonal IgG and free LAMBDA light chain spike which is compatible with multiple myeloma. Ocular manifestations of multiple myeloma are rare and adverse, hence there is no single comprehensive protocol that can be extrapolate to these various dilemmas.

Case Presentation
A 50 year-old woman presented to the hospital with a complaint of bilateral gradual progressive visual loss for the preceding three years, associated with cloudy discoloration of cornea in both eyes (Figure 1). Best-corrected visual acuity was 15/20 in both eyes. Pupillary examination and ocular movement were normal.

Slit-lamp examination showed copper deposition (brown/green), diffusely scattered in the Descemet's membrane and deep layers of central cornea along with anterior lens capsule. Apart from this, she had no remarkable past medical history, familial history or exposure to copper. Laboratory evaluations demonstrated hypercupremia, normal ceruloplasmin levels and rose. ESR: 50 (M: up to 12 mm/hr and F: up to 22 mm/hr)

Upon further assessment, there were not any evidence of distress and she appeared to rest in a comfortable state. Moreover, her vital signs were well within normal limits. Except for the ocular changes, physical examinations were unremarkable with no hepatosplenomegaly or palpable lymphadenopathy.

Skull X ray of AP and lateral view showed multiple punched out lesions over skull bone (Figure 2).

In the light of findings through the pattern of copper deposition as previous reported, we decided to evaluate the patient for suspected plasma cell disorders. The patient underwent a serum protein electrophoresis, which showed a monoclonal IgG (25.020. nl range: 6.57-18.37) and free LAMBDA light chain spike (42.1nl range: 5.7-26).

Bone marrow aspiration and biopsy revealed an increased population of abnormal and dysplastic plasma cell (35% of the cellular elements) which was compatible with the diagnosis of multiple myeloma. The patient was diagnosed with multiple myeloma based on the laboratory, histology and imaging findings. The clinical finding of copper deposition in Descemet's membrane and in the anterior lens capsule associated with in the Descemet's membrane and deep layers of central cornea along with anterior lens capsule are consistent with the previous reported patients with multiple myeloma [1-5] and monoclonal gammopathy of undetermined significance (MGUS) [6-10]. Although in this case the posterior lens capsule remains intact. The reason behind ocular deposition of copper in multiple myeloma and its unique pattern which differs from Wilson disease still eludes us. One suggested mechanism for this phenomenon is that IgG λ has an affinity of copper in multiple myeloma and consequently contribute to the copper transfer into the anterior chamber and then diffuse and bind to Descemet's membrane [4]. Copper has a special affinity for the basement membrane and accumulations in Descemet's membrane as we can see the characteristic
The patient was treated with Velzomib, dexamethasone and thalidomide (so called VTD regimen). Moreover D-penicillamin was added to decrease serum copper level. After completed remission she was referred to a center for autologous Bone Marrow Transplantation.

References