



Bilateral Posterior Scleritis in a Case of Multiple Myeloma

Multipl Myelomlu Bir Olguda Bilateral Arka Sklerit

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Summary

A 78-year-old female patient presented with complaints of decreased vision accompanied by redness and pain in both eyes for 1 week. She was diagnosed as having immunoglobulin G kappa type smoldering multiple myeloma 1 year ago. The dilated fundus examination revealed choroidal folds at the posterior pole and exudative retinal detachment in the peripheral area in both eyes. According to these findings, the patient was diagnosed with bilateral posterior scleritis and corticosteroid treatment was started. Osteoporotic bone lesions, anemia, and hypercalcemia were detected during the systemic examination, and the patient was diagnosed with immunoglobulin G kappa type multiple myeloma. (Turk J Ophthalmol 2014; 44: 481-3)

Key Words: Multiple myeloma, smoldering multiple myeloma, posterior scleritis

Özet

Yetmiş sekiz yaşında kadın hasta her iki gözünde 1 haftadır devam eden görmeye azalma, kırmızılık ve ağrıyla başvurdu. Hastaya 1 yıl önce immünglobülin G kappa tipi sessiz multipl myelom tanısı konulmuştu. Dilate fundus muayenesinde her iki gözde arka kutupta koroid kıvrımları ve perifer bölgede eksüdatif retina dekolmanı mevcuttu. Bu bulgular ile hastaya bilateral arka sklerit tanısı konuldu ve kortikosteroid tedavisi başlandı. Sistemik muayenede osteoporotik kemik lezyonları, anemi ve hiperkalsemi saptandı ve hastaya IgG kappa tipi multipl myelom tanısı konuldu. (Turk J Ophthalmol 2014; 44: 481-3)

Anahtar Kelimeler: Multipl myelom, sessiz multipl myelom, arka sklerit

Introduction

Multiple myeloma (MM) involves the cells of the immune system and is the third most prevalent blood cancer after non-Hodgkin's lymphoma and chronic lymphocytic leukemia. The majority of patients are elderly, with only 3.8% of affecting patients younger than 45.¹ Smoldering (asymptomatic) multiple myeloma (SMM) is an asymptomatic plasma cell proliferative disorder associated with high risk of progression to symptomatic MM or amyloidosis. In 2003, the International Myeloma Working Group agreed on a definition of SMM consisting of a serum M-protein of ≥ 3 g/dL and/or $\geq 10\%$ bone marrow plasma cells with no evidence of end-organ damage.² Ocular involvement of MM is rare, but it can occur in the orbit, extraocular muscles, and within the uveal tract.³

Posterior scleritis is seen commonly in middle-aged females, associated with mild ocular congestion, globe tenderness, and varying amount of vision loss. Posterior scleritis can occur in various ways mimicking orbital tumors, orbital inflammation, optic neuritis, and vasculitis.⁴

The aim of the study was to report a case with MM presented with bilateral posterior scleritis.

Case Report

A 78-year old woman presented with complaints of decreased vision accompanied with redness and pain in both eyes for 1 week. A detailed history ruled out ocular trauma or surgeries or similar episodes in the past in either eye. Immunoglobulin G (IgG) kappa type SMM was diagnosed 1 year ago. At presentation, the best-corrected visual acuity

in right eye was 20/40 and in left eye 20/30. Intraocular pressure was 17 mmHg in right eye and 14 mmHg in left eye. Ocular movements were free and painful in both eyes. The bulbar conjunctiva showed ciliary congestion in both eyes. The dilated fundus examination revealed choroidal folds at the posterior pole in the right eye and an exudative retinal detachment in the peripheral area in both eyes (Figure 1). The ultrasounds B scan demonstrated a large hypoechoic area in the sub-Tenon's space (T-sign) suggestive of periocular fluid collection and thickened sclero-choroidal complex (Figure 2). Orbital ultrasound did not show evidence of any tumor or any increase in extraocular muscle thickness. Fundus fluorescein angiography showed few areas of pinpoint hyperfluorescence in the early phase with leakage in the late phase, leakage from the optic disc and pooling of dye in areas of exudative detachment in the late phases in both eyes (Figure 3). These findings led to bilateral posterior scleritis diagnosis. The patient was consulted with rheumatology and hematology department, a complete blood count, erythrocyte sedimentation rate, antinuclear antibodies, rheumatoid factor, C-reactive protein, chest X-ray, and sacroiliac joints X-ray were done. Osteoporotic bone lesions and hypercalcemia were detected during the systemic examination and IgG kappa type MM was diagnosed. Intravenous pulse methyl prednisolone 1000 mg daily for 3 days was started and followed by oral prednisolone 60 mg (1 mg/kg bodyweight) tapered over the next month. After 1 month, the ciliary congestion was significantly reduced and exudative detachment and choroidal folds regressed. The patient refused additional chemotherapy. After 1.5 years, the patient had similar posterior scleritis attack and treated with same steroid regimen. The patient was consulted with the hematology department again and treated with chemotherapy (melphalan and prednisone). Throughout the 7 years, the patient has been under hematologic and ophthalmologic control and has never experienced an ocular attack or a new organ involvement during follow-up.

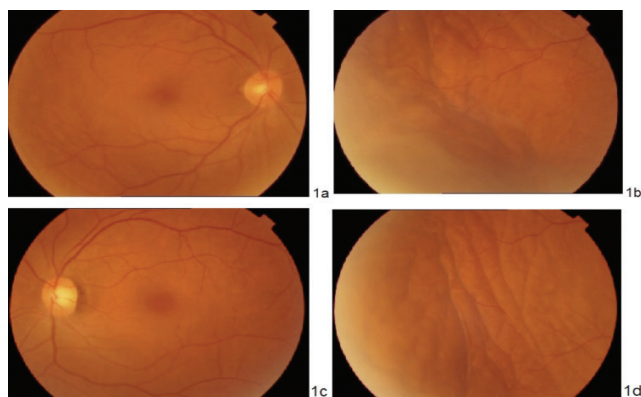


Figure 1. The dilated fundus examination revealed choroidal folds at the posterior pole (a) and an exudative retinal detachment in the peripheral area (b) in right eye. Posterior pole was normal in the left eye (c), but peripheral exudative retinal detachment was observed (d)

Discussion

Multiple myeloma is a progressive and incurable but treatable cancer, resulting in uncontrolled proliferation of neoplastic plasma cells in the bone marrow and excessive production of monoclonal immunoglobulins. Myeloma is classified by the type of immunoglobulin that is abnormally produced. The most common type is IgG, followed by IgA, IgM, IgD and IgE with respect to frequency of occurrence. Osteoporotic bone lesions, anemia, kidney disease, bone marrow failure, hypercalcemia and susceptibility to infection are hallmarks of the disease.⁵

Ocular involvement with myeloma is uncommon, but can be the first manifestation of the disease and can appear in almost every ocular structure. IgG type multiple myeloma, whether lambda or kappa light chain, is reported to be a risk factor for orbital involvement.³ Orbital, conjunctival, uveal, lacrimal sac,



Figure 2. The ultrasound B scan showed a large hypoechoic area in the sub-Tenon's space (T-sign) suggestive of periocular fluid collection and thickened sclero-choroidal complex

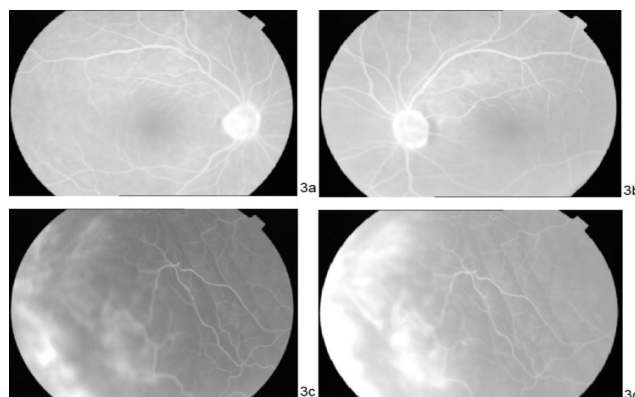


Figure 3. Fundus fluorescein angiography showed staining of the optic disc in the late phase in the both eyes (a, b) and few areas of pinpoint hyperfluorescence in the early phase with leakage in the late phase, and pooling of dye in areas of exudative detachment in the late phases in the both eyes (c, d)

and lacrimal gland infiltration have been described. Ocular manifestations also range from chorioretinopathy (e.g., choroidal infiltrate, choroidal effusion, retinal capillary microaneurysms, hyperviscosity), neuro-ophthalmic abnormalities (e.g., extraocular muscle paresis or direct myelomatous infiltrate, papilledema), and corneal deposition (crystal or copper) to opportunistic infections of ocular structures.⁶⁻⁹ Until now there was no case of co-existence of multiple myeloma and scleritis or scleral involvement in the literature. No solid tumor such as plasmacytoma was observed in the patient and we could not execute a biopsy.

Multiple myeloma is usually treated with systemic chemotherapy and bone-marrow transplantation for the blood and marrow involvement.⁵ Our patient was treated chemotherapy for systemic disease along with steroid therapy during the posterior scleritis attacks.

In our study, we reported a 78 year-old woman with MM who was presented with bilateral posterior scleritis. The findings regressed with steroid therapy but recurred after 1.5 years. After second attack, the patient was treated with chemotherapy. We think that the posterior scleritis is associated with the MM because the patient had two attacks during the activation of the MM. After the chemotherapy both the MM and ocular findings are under control, the patient never experienced an ocular attack or new organ involvement during follow-up.

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