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A Rare Presentation Of Paraparesis Due To Epidural Myeloma With Neuropathic Onset

Case Report

Epidural Myeloma bağlı Nöropatik başlangıçlı nadir bir Paraparezi Olgusu

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ABSTRACT

Multiple myeloma is a plasma cell dyscrasias that often complicating with neurological symptoms. Among these vertebral fractures, spinal canal compromises and peripheral neuropathies are best known. In this report, we present a case which peripheral neuropathy precedes acute paraparesis that is seen very rarely. A 58-year-old woman was admitted to our hospital with fatigue, nonspecific back pain and numbness in her legs. During hospitalisation she was treated for peripheral neuropathy that acute paraparesis was developed later on. Urgent lumbar magnetic resonance imaging showed an epidural mass that excised totally via dorsal midline approach. Pathology confirmed the diagnosis of plasmacytoma. Patient was able to mobilize independently one month after discharge.

Key words: *Myeloma, Neuropathy, Plasmacytoma*

ÖZET

Multiple myeloma sık nörolojik komplikasyonların izlendiği bir plazma hücreli kan hastalığıdır. Bu komplikasyonlar arasında periferik nöropatiler, spinal kanal basıları ve omurga kırıkları en iyi bilinenlerdir. Bu çalışmamızda, periferik nöropatinin akut paraparezinin öncüsü olduğu bir hastayı sunduk. 58 yaşında kadın hasta yorgunluk, nonspesifik bel ağrısı ve bacaklarda uyuşma şikayeti ile hastanemize başvurmuş. Periferik nöropati tanısıyla tedavisi başlanan hastada daha sonra akut paraparezi gelişmiş. Çekilen acil lomber MRG'de epidural kitle izlenen hasta opere edilerek kitle total olarak çıkartıldı. Patoloji sonucu plazmasitom olarak gelen hasta 1 ay sonra mobilize olarak taburcu edildi.

Anahtar Kelimeler: *Miyelom,Nöropati, Plazmasitom*

INTRODUCTION

Multiple myeloma is a generalized disease of malignant plasma cells (1). It is the most frequent primary tumour of the spine typically located in the vertebral body of the lower thoracic, lumbar and then cervical spine (14). Therefore, back pain is the common presenting symptom and sometimes root compression cause severe sciatalgia like disc herniation (12,15). Besides these findings, peripheral neuropathy is another presentation (13). Also, because of the vertebral collapse and epidural invasiveness, spinal cord compression could be developed in the patients (12). In this paper; we present a case of compression myelopathy due to an epidural mass of IgA myeloma which initial presenting symptom was numbness in lower extremities that diagnosed as peripheral neuropathy.

CASE REPORT

A 58-year-old woman was admitted to our hospital with fatigue and nonspecific back pain. She had a history of anemia, which is treated inadequtely. At emergency room her hemoglobin level was 9,9g/dl. She was hospitalized by internal medicine for investigation of anemia. The day after hospitalization she felt numbness in her legs. Neurological examination revealed hypoesthesia in distal lower extremities. Electroneuromyography (ENMG) was displayed sensorial type neuropathy and Gabapentin 600 mg was given 3x1 daily. Lumbar anteroposterior X-Ray graphy showed only degenerative changes and analgesic treatment was given meanwhile. Acute paraparesis was developed later on. Neurological examination revealed paraperesis (4/5 in proximal lower limb and 3/5 in distal lower limb). Urgent lumbar and thoracic magnetic resonance imaging (MRI) was displayed an epidural mass epidural mass at the level of T11-12 and L2-3 (Figure-1). Patient was operated via dorsal midline approach immediately. The mass was located epidurally that is very soft and easily aspirated and excised totally. Histopathological result was plasma cell tumour (Figure-2). After then, serum immunoelectrophoresis revealed the monoclonal component of immunoglobulin A (IgA) protein and kappa-light chain. IgA level of serum was 38,7 g/L (0,7-4). No Bence-Jones proteinuria was detected. Postoperative course was uneventful and the patient's symptoms gradually improved. Systemic chemotherapy was started and radiotherapy was planned after wound healing.

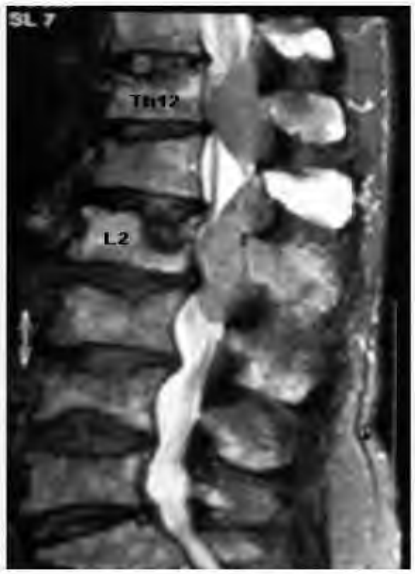

Figure 1: *T2-Weighted sagittal lumbar MRI shows intraspinal mass lesion located at T12 and L2 level*

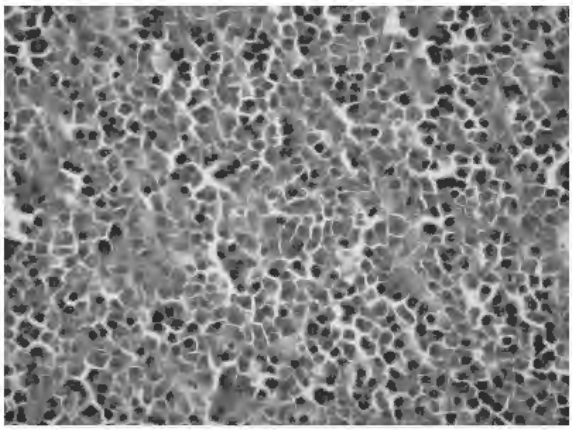

Figure-2: *Histopathological examination of the specimen revealed plasmocytoid tumoral cells with marked nuclear atypia and invasion of bone marrow with tumoral cells. (400x, H&E)*

DISCUSSION

Multiple myeloma is a primary neoplasm of plasma cells, which proliferated malignantly. It affects males slightly more than females; the male: female ratio is 1,6:1,0. The annual incidence is between 5 and 10 per100.000 population (10). Despite the disease occurs mostly in elderly and median age is 66 year, patients younger than 40 year are affected 2% (5). Patologic secretion of a monoclonal paraprotein could be documented in almost all patients. Amog these IgG secreting myeloma is the most common, IgA and the others with respectively 52% and 21%(11). Because of the localization of the hematopoietic cells, it affects mostly axial skeleton. Thoracic region and then lumbar and cervical region is affected in decreasing order (3). As a result of this localization the main symptom is usually back pain in 80% of patients. Also fatigue is another presentation symptom. Mild anemia and hypercalcemia that responsible for the renal failure is seen 30% of patients (1,5). Destruction of vertebral bodies and expension of tumors beyond the cortex as epidural mass may result in spinal cord compression. Woo et al reports the case of spinal cord compression due to epidural mass 15 % and fracture 8 % of 97 multiple myeloma patients (15). Lecouvet et al reported that 55 to 70 % of multiple myeloma patients are affected from compression fracture with or without spinal cord compression (7). However compression due to an epidural mass is seen less than fractures and reported with an incidence of 10-20 % (12). Beside these, another neurological impairement, which is seen in myeloma disease, is neuropathy also. Walsh et al found neuropathy clinically 13% and electrophysiologically 39% in a prospective study (13). Despite neuropathy is mostly sensorimotor type, purely sensory type may occur infrequently. Generally neuropathy is presented in gradual onset but seldom acute or subacute onset may be seen like in presented case (2,4). In pathogenesis autoantibody activity of immunoglobulins against myelin sheath is responsible in this process. Despite this condition is more pronounced in IgM monoclonal gammopathy (MG), also IgG or IgA MG's are associated with neuropathy (6). Maggy et al reviewed fifteen IgA/IgG MG patients and reported that sensorial neuropathy is prominent in IgM MG but in IgA/IgG MG, clinic is more heterogeneous and he suggested a different pathomechanism also (8). For exact diagnosis, serological investigations such as immunoelectrophoresis, serum immunoglobulin level and urinary bence-jones proteins must be done. Plain radiograms may show the vertebral fractures but not able to show the soft tissue masses (15). For that reason magnetic resonance imaging of axial skeleton must be done for differential diagnosis and planning surgery. Currently first choice of treatment is radiotherapy and/or chemotherapy. Although, these methods successfully control the disease and the pain, in the case of neurological impairment, spinal instability and intractable pain surgery is indicated (3). However, treatment of the neuropathy needs removing the monoclonal B cells and the circulating autoantibodies (6). Mankodi et al reported such a case that successful treatment of the disease controls the symptoms of neuropathy (9).

We concluded that even in the presence of a normal or degenerative X-Ray and diagnosis of peripheral neuropathy in myeloma, we couldn't exclude the presence of an epidural mass. Because the treatment modalities are strictly different in neuropathy and epidural myeloma, in case of epidural compression, any delay in diagnosis may lead to neurological compromise. For that reason, we should keep in mind that sensorial or motor neuropathies associated with myeloma may precede a threatening case. So, for exact differentiation spinal MRI should be added to the serological tests and in the case of neurological compromise urgent surgical decompression should be carried out.

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