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Case report
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A rare case of newly diagnosed multiple myeloma presenting to the emergency department with acute paraplegias

Multipl myelom'un acil servise prezentasyonu

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Abstract

Multiple myeloma (MM), neoplastic proliferation of monoclonal plasma cells in bone marrow, constitutes 1% of cancers. We discuss a rare case of newly diagnosed MM presenting to the emergency department with sudden strength loss in both legs and inability to walk, together with the current literature. Impaired balance and 2/5 strength loss in the bilateral lower extremities was determined in a 42-year-old man presenting due to weakness in both legs and inability to walk, noticed on waking in the morning. At blood tests, uric acid was 8.4 mg/dl, total protein 98.8 g/L, globulin 60.8 g/L. Magnetic resonance imaging revealed a mass between T_2 and T_4 vertebrae. At mass biopsy immunohistochemical investigation, lesion cluster of differentiation (CD)38, CD138, endomysial antibody, kappa and lambda cells exhibited immune reactions. Serum IgA was 0.27 g/L, IgM 0.28 g/L, IgG 44.36 g/L, beta-2 microglobulin 4.78 mg/L, lambda light chain 0.230 g/L, and kappa light chain 5.25 g/L. Mass biopsy revealed plasma cell tumor, with atypical plasma cells at 52%, which was compatible with MM at bone marrow aspiration biopsy. Neoplastic plasma cell proliferation should be considered for differential diagnosis in sudden strength loss in the lower extremities.
Keywords: Emergency medicine, Multiple myeloma, Plasmacytoma, Acute paraplegia

Öz

Kemik iliğinde monoklonal plazma hücrelerinin neoplastik proliferasyonu olan multipl myelom (MM), tüm kanserlerin %1'ini oluşturur. Bu yazıda ani gelişen her iki bacakta kuvvet kaybı ve yürüyememe şikayeti ile acil servise müracaat eden hastada tespit edilen ve nadir görülen yeni tanı MM literatür eşliğinde tartışmayı amaçladık. Sabah fark ettiği her iki bacakta kuvvet kaybı ve yürüyememe şikayeti ile acil servise başvuran 42 yaşında erkek hastanın alt ekstremitelerinde 2/5 kuvvet kaybı ile beraber dengesini sağlamada bozukluk tespit edildi. Kan tetkiklerinde ürik asit 8,4 mg/dl, total protein 98,8 g/L, globülin 60,8 g/L olarak tespit edildi. Manyetik rezonans görüntülemede torakal $(T)_2$-T_4 vertebralar arasında kitle izlendi. Kitle biyopsisi immünohistokimyasal çalışmada lezyona ait hücreler cluster of differentiation (CD)38, CD138, endomysial antibody, kappa ve lambda ile immün reaksiyon gösterdiği gözlendi. Serum IgA 0,27 g/L, IgM 0,28 g/L, IgG 44,36 g/L, beta-2 mikroglobulin 4,78 mg/L, lambda hafif zincir 0,230 g/L, kappa hafif zincir 5,25 g/L olduğu tespit edildi. Kitlenin biyopsi incelemesi plazma hücreli tümör, kemik iliği aspirasyon biyopsisi ise MM ile uyumlu ve atipik plazma hücreleri %52 olduğu tespit edildi. Hastada MM düşünüldü. Alt ekstremitelerde ani gelişen kuvvet kaybı şikayetiyle acil servise müracaat eden hastalarda ayırıcı tanıda plazma hücrelerinin neoplastik proliferasyonu olabileceği düşünülmelidir.
Anahtar kelimeler: Acil tıp, Multipl miyelom, Plazmasitom, Akut parapleji

Introduction

Multiple myeloma (MM), the neoplastic proliferation of monoclonal plasma cells in bone marrow, constitutes 1% of all cancers and approximately 10% of hematological malignancies. The most common clinical presentation is bone pain, while pathological fractures, recurrent bacterial infections, and kidney failure are less common. Plasmacytoma is a solitary neoplasm of monoclonal plasma cells [1,2]. Clinical findings associated with spinal cord or nerve-root compression are observed, depending on the location [3]. This report discusses a case of plasmacytoma associated with rare MM determined in a patient presenting to the emergency department with acute onset loss of strength in the bilateral lower extremities and inability to walk, in the light of the current literature.

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Case presentation

A 42-year-old man, from whom verbal consent for this case report was obtained, presented to the emergency department due to loss of strength in the bilateral lower extremities and inability to walk, which he noticed on waking in the morning. On arrival, the patient's general condition was average, he was lucid and cooperative, and vital signs were normal. No history of disease or trauma was present. He described a sensation of numbness in the bilateral plantar regions at night, followed by onset of pain in the bilateral proximal of the lower extremities, but he was able to walk with support. By morning he reported becoming unable to walk or to support his body weight in a standing position. He also reported that the feeling of numbness in the soles experienced at night had spread as far as the umbilical region. The patient reported no urine or fecal incontinence. Physical examination revealed 2/5 loss of strength in the bilateral lower extremities and impaired balance, as if the patient were about to fall forward from a standing position. Deep tendon reflexes were hyperactive in the bilateral lower extremities. At blood tests, uric acid was 8.4 mg/dl (range 3.5-7.2), creatinine 0.99 mg/dl (range 0.67-1.17), calcium (Ca^{++}) 9.9 mg/dl (range 8.4-10.5), potassium (K^{+}) 3.7 mEq/L (range 3.5-5.1), total protein 98.8 g/L (range 66-83), albumin 38 g/L (range 35-52), globulin 60.8 g/L (range 15-37), lactate dehydrogenase (LDH) 285 U/L (range <247), C-reactive protein (CRP) 27 mg/L (range 0-5), sedimentation 38 mm/hour (range 0-30), and hemoglobin 13.6 g/dl (range 14.1-18.1). No pathology was determined in the cranial computerized tomography (CT) or magnetic resonance imaging (MRI). Spinal CT revealed extensive medullary lytic lesions in the cervical $(C)_7$, and thoracic $(T)_{2,4, \text{ and } 11}$ vertebrae, in all lumbar vertebrae, in the bilateral ileac bones and the sacrum. Spinal MRI revealed a mass lesion, T1 hypointense and T2 mildly hyperintense, with contrast involvement, 5 cm in length and with an anteroposterior diameter of 9.2 mm, compressing the cord from the right posterolateral to the anterior region, with an extra-axial location in the right half of the posterior spinal canal at the T_2 vertebra level, extending inferiorly to the T_4 vertebra level. Nodular lesions, T1 hypointense and T2 hyperintense, and exhibiting contrast involvement, were observed in the $T_{10,11}$ vertebral corpi (Figure 1). The patient was transferred to the neurosurgery department for surgery. Biopsy material was collected with excision of the thoracic intradural extramedullary mass and sent to the pathology laboratory. The relation with MM was investigated in consultation with the hematology department. Peripheral smear result was reportedly normal. At immunohistochemical study, cells from the lesion exhibited cluster of differentiation (CD) 38, CD138, endomysial antibody (EMA), and immune reaction with kappa and lambda. Serum IgA was 0.27 g/L (range 0.7-4), IgM 0.28 g/L (range 0.4-2.3), IgG 44.36 g/L (range 1-16), beta-2 microglobulin 4.78 mg/L (range 0,8-2,4), serum free kappa light chain 5.25 g/L (range 1,7-3,7), and serum free lambda light chain 0.230 g/L (range 0.9-2.1). Lambda light chain in 24-h urine was <3.720 mg/L (range <10), and the kappa light chain value was <6.690 mg/L (range <15). The mass biopsy was reported as an IgG/Kappa monotypic plasma cell tumor. Bone marrow aspiration was compatible with MM, and atypical plasma cells were determined at 52% (Figure 2). Plasmacytosis arising from MM was suspected, and the patient was discharged with polyclinic treatment and follow-up.

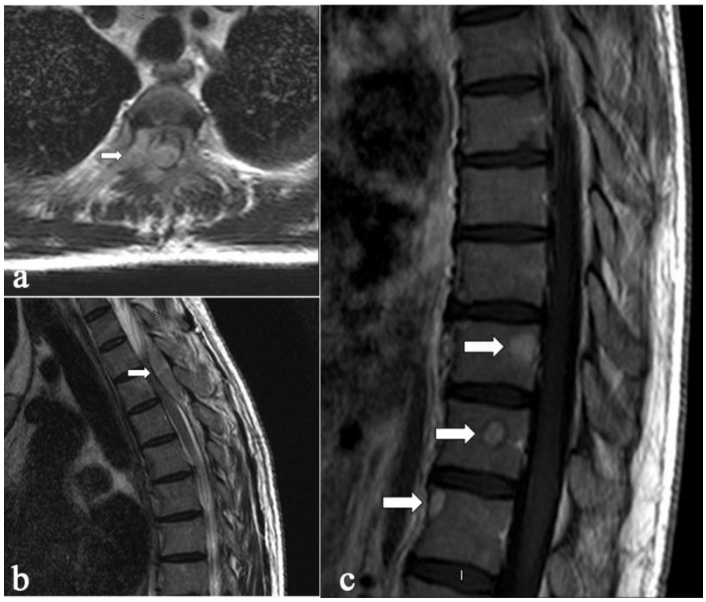

Figure 1: Plasmocytoma transverse (a), sagittal (b) and vertebral nodular lesion (c) images at MRI

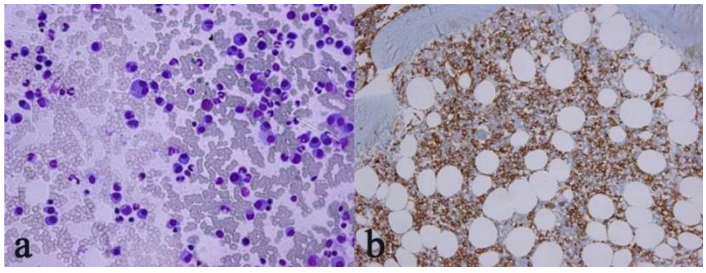

Figure 2: a) Plasma cells in bone marrow aspiration biopsy preparate (Giemsa X400) and b) IgG involvement (immunohistochemical X400)

Discussion

Plasma cell tumors arise from B-lymphocytes as a result of the proliferation and accumulation of plasma cells that synthesize immunoglobulin [4]. These include MM, monoclonal gammopathy of undetermined significance, plasma cell leukemia, solitary plasmacytoma, Waldenström macroglobulinemia, primary amyloidosis (AL), heavy chain disease, POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, M Protein, and Skin Changes) syndrome, Type I and II cryoglobulinemia, and light chain deposition disease [5,6]. MM is a lymphoproliferative disease that causes malignant proliferation of immunoglobulin-secreting plasma cells which most commonly involves bone marrow (42%). The annual incidence in the USA is 3-4/100,000 [7]. Although not enough is known about the etiology, chronic exposure to viral agents and radiation has been implicated [8]. No etiological cause was determined in our patient.

The most commonly involved bones in MM are the vertebrae in particular, along with the calvarium, pelvis, rib, scapula, humerus and femur [9,10]. Diffuse involvement was observed in the vertebral column in our patient. The most common findings in MM are osteolytic bone lesions, osteoporosis, and resultant pain. Anemia-related symptoms and findings, infections resulting from immune paralysis, a disposition to hemorrhage due to various types of hemostasis impairment, polyneuropathy, paraplegia and quadriplegia

resulting from vertebral fractures, root compression symptoms, symptoms of hyperviscosity syndrome, and hyperuricemia may also be seen [11]. Despite the absence of back pain, we attributed the acute onset pain and strength loss in the bilateral lower extremities to spinal cord compression due to spinal plasmacytosis. Hyperuricemia was also present, but no bone fracture was determined.

Direct radiography is still the primary diagnostic technique in determining destructive osseous lesions in the diagnosis of MM. Small, distinct lytic lesions with well-demarcated margins, and staple-like holes are observed at radiography. CT is a more sensitive diagnostic technique in showing osseous lesions. Lytic lesions at CT exhibit expansile masses with a soft tissue component, diffuse osteopenic bone fractures, and osteosclerosis. MRI assists CT in terms of mass staging and spread [12]. No pathology was observed at x-ray in our case, while CT revealed medullar lytic lesions, and MRI revealed a spinal mass and nodular lesions.

Plasmacytoma constitutes 3% of plasma cell disorders. Clinical findings vary, depending on the location, while laboratory findings are similar to those of MM. However, prognosis is better in comparison to MM [13]. It frequently involves the thoracolumbar region, and particularly the vertebral body [14]. The lesion arises from regions of bone marrow function. Diagnosis is confirmed by tissue biopsy in addition to clinical and laboratory findings, as in the present case [15].

With the spread of plasma cells in bone marrow caused by myeloma, it restricts erythropoiesis, while generally normocytic normochromic anemia or rarely macrocytic or microcytic anemia occur as a result of obstruction of erythropoiesis by means of release of interleukin-6 by bone marrow stromal and endothelial cells. Hypercalcemia is an important finding of diagnostic value in myeloma, measures the tumor burden and indicates organ damage, and is seen in 18-30% of cases. Increases in creatinine values exceeding 2 mg/dl are seen in 25% of patients with MM at time of diagnosis. In other words, kidney failure of varying degrees of severity is present in approximately half of patients. Serum LDL elevation is seen in between 7% and 11% of newly diagnosed MM cases. Myeloma plasma cells exhibit CD 38-, CD 138-, and CD 79a-positivity [15]. Mild, asymptomatic anemia was present in our patient. No hypercalcemia or kidney failure were observed, but LDL levels were elevated. Myeloma plasma cells also exhibited immune reaction with CD38 and CD138.

Diagnostic criteria for MM were revised by the International Myeloma Working Group (IMWG) in 2014, and were defined as clonal bone marrow plasma cells ≥10% or biopsy-proven bony or extramedullary and serum M protein (monoclonal immunoglobulin) IgG ≥3 g/dL, IgA >1 g/dL and presence of a myeloma-defining event, in other words, one or more CRAB features or findings (elevated serum calcium level, renal failure, anemia, and bone lesions) or one or more SLiM criteria (clonal bone marrow plasma >60%, involved/uninvolved serum-free light chain ratio >100, presence of more than one focal lesion 5 mm in size or larger at whole-body MRI) [16]. Our patient was assessed as stage 2 according to the International Staging System.

In addition to various classic chromosome analysis tests, structural changes in chromosomes, which are related to risk levels in MM patients, can be determined using molecular cytogenetic tests such as polymerase chain reaction and fluorescent in situ hybridization. Chemotherapy and autologous stem cell transplantation are performed in the treatment of MM. Prognosis depends on various factors, but is generally poor [16].

Conclusion

The possibility of neoplastic proliferation of plasma cells should be considered for differential diagnosis in patients presenting to the emergency department with sudden onset loss of strength in the lower extremities and no history of trauma.

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