

A Rare Emergency Condition in Neurosurgery: Foot Drop Due to Paget's Disease

Nöroşirürjide Nadir Bir Acil Durum: Paget Hastalığına Bağlı Düşük Ayak

ABSTRACT

Paget's disease is a chronic, focal skeletal disorder that usually affects the pelvis and spine. Spinal cases are generally asymptomatic; in the symptomatic cases, the neurological dysfunctions are related to non-compressive vascular defects, hemorrhage, sarcomatoid degeneration, spinal stenosis, or pathological fractures, primarily in the lumbar region. The Neurosurgeon should have a fundamental understanding of the complications of Paget's disease and should be familiar with the indications for treatment, as well as available medical and surgical therapies. In the present paper, we report a case of Paget's disease that presented with an isolated foot drop due to a pathological fracture of L5 vertebra, and then discuss the therapeutic strategies presented in the literature.

KEYWORDS: Foot drop, Lumbar vertebrae, Paget's disease, Pathological fracture

ÖZ

Paget hastalığı sıklıkla pelvis ve omurgayı tutan bir kronik, fokal iskelet rahatsızlığıdır. Spinal olgular genelde asemptomatik olmakla birlikte, semptomatik olgularda nörolojik disfonksiyonlar öncelikle lumbar bölgedeki nonkompresif vasküler defektler, hemoraji, sarkomatoid dejenerasyon, spinal dar kanal veya patolojik fraktürler ile ilişkilidir. Bir beyin cerrahı, Paget hastalığının mevcut cerrahi ve tıbbi tedavilerini, endikasyonlarını bilmesi gerektiği kadar hastalığın komplikasyonlarında farkında olması gereklidir. L5 vertebraesindeki patolojik fraktüre bağlı izole düşük ayakla sonuçlanmış bir Paget hastalığı olgusu, literatürdeki tedavi stratejileri ile birlikte tartışılmıştır.

ANAHTAR SÖZCÜKLER: Düşük ayak, Lumbar vertebra, Paget hastalığı, Patolojik fraktür

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INTRODUCTION

Paget's disease is characterized by excessive bone formation with resorption and abnormal remodeling. Common sites of involvement include the spine, pelvis, skull, femur, and tibia. The disease is polyostotic in approximately 66% of the cases, and half of these patients have a lesion in the vertebral column (9).

Although spinal Paget's disease is usually asymptomatic, common symptoms encountered in clinical practice are back pain, numbness and tingling of the feet, difficulty walking, paralysis of the legs, and abnormal bladder and bowel function. We are not aware of a reported case of spinal Paget's disease that presented with a foot drop due to pathological fracture of a vertebra.

CASE REPORT

A 38-year-old male patient was admitted to our clinic with complaints of sudden pain in the lower back, as well as numbness and weakness in his left foot. On neurological examination, he had a foot drop in his left leg and sensory loss in the left L5 dermatome. Magnetic resonance imaging (MRI) revealed diffuse medullar expansion with a rough trabecular pattern on the corpus and the posterior components of the L3 vertebra, causing relative spinal canal stenosis (Figure 1). There was an osseous fragment located in the posterior paramedian part of the L5 vertebral corpus, compressing the left L5 nerve root. The lesion was hyper-intense on T2-weighted sequences, and mixed-intense on T1-weighted images (Figure 2A). The MRI also showed a heterogeneous hyper-intensity on the left side of the L5 vertebral corpus, and cortical loss due to an oblique fracture at the left posterior paramedian region of the corpus (Figure 2A,B). Together these findings suggested Paget's disease or hemangioma. We



Figure 1: T2-weighted sagittal sequence of magnetic resonance imaging showing diffuse pagetic involvement in L3 vertebra and partial involvement in L5 vertebra.

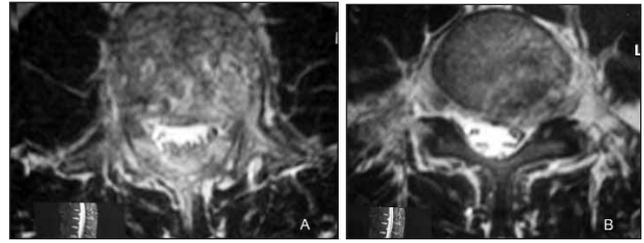


Figure 2: A: Magnetic resonance imaging showing diffuse medullar expansion of the L3 vertebra and relative spinal canal stenosis. B: Magnetic resonance imaging showing severe root compression on the left L5 nerve. Note heterogeneous hyperintensity on the left side of the L5 vertebral corpus and prominent foraminal stenosis due to the retropulsed osseous fragment.

performed a left L4 partial hemilaminectomy and left L5 foraminal decompression. Histopathological examination showed irregular, disorganized bone lamellas with scattered, dissociated, plump Paget cells (Figure 3A). The presence of hyper-cellular bone trabecules in a woven pattern confirmed the diagnosis of Paget's disease (Figure 3B). Bone scintigraphy revealed an increase of the activity index in L1 and L5 vertebrae and the sternum. The patient's serum alkaline phosphatase level was 418 U/L (N: 70-290 U/L). We began treatment with daily Etidronate Na (400 mg, orally). The postoperative course of the patient was uneventful, and he was discharged from the hospital 2 days postoperatively. The treatment response was good, and alkaline phosphatase levels returned to nearly normal. His symptoms quickly improved and he had no neurological deficits 6 months after the operation.

DISCUSSION

Paget's disease is a chronic disorder of the bone that presents as enlarged or deformed bones at one or more skeletal sites. The underlying pathophysiological

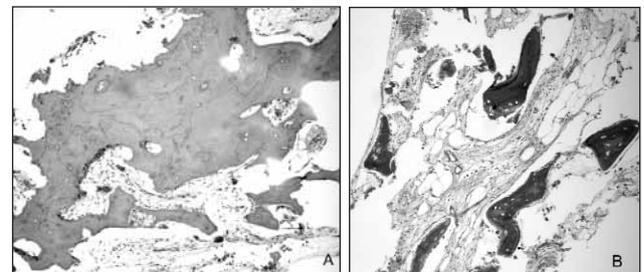


Figure 3: A: Photomicrograph shows osteoblastic and osteoclastic activities with irregular, disorganized bone lamellas (H&E, original magnification x100). B: A photomicrograph of surgical material revealing woven bone pattern that confirms an active pagetic lesion (Trichrome stain, original magnification x100).

mechanisms of this disease are still unclear, but include genetic factors and paramyxovirus infection (5,7).

Paget's disease is generally polyostotic and asymmetric. The pelvis and spine are the most common sites of involvement. The distribution of Paget's disease in the spine is similar to that of osteoarthritis (2). Meunier and colleagues (8) assessed the whole body skeletal scintigrams of 170 untreated pagetic patients, and found involvement in the lumbar (58%), thoracic (45%) and cervical regions (14%). Among lumbar vertebrae, the middle ones were the most commonly affected, whereas L5 was the least commonly affected (6). In the present case, we observed total pagetic involvement in L3 vertebra and partial involvement in L5 vertebrae, as well as the sternum.

Spinal Paget's disease manifests with cortical thickening encasing the vertebral margins, which gives rise to the "picture frame" appearance on radiographs. MRI and CT demonstrate enlarged bones with trabecular coarsening and increased cortical thickness. Radionuclide bone scans are more sensitive than radiographs for the diagnosis of Paget's disease. Bone scintigraphy demonstrates increased radionuclide uptake in the region of abnormal bone in Paget's disease (3,9,11).

Spinal Paget's disease is usually asymptomatic (95%-97%), with back pain the most common presenting symptom (3). Neurological dysfunction results from spinal stenosis, pathological fracture, hemorrhage, sarcomatous degeneration, or non-compressive vascular deficiencies (6,3,4,10,11). Pathologic fractures in these cases generally occur in the lumbar region (1). In the present case, a previously asymptomatic patient presented with a foot drop due to a pathological fracture of the L5 vertebra. As in this case, a pathological fracture may occur even at a partially involved vertebra. Fractures can be seen at any location with higher stress and degree of involvement, as in our case. This is the first report of foot drop due to a pathological fracture in Paget's disease.

Pagetic spinal stenosis requires symptomatic treatment with antipagetic drugs (12). Decompressive surgery may be required when resistance to medical therapy or serious neurological deficits are present. Surgery is indicated as a primary treatment with neural compression due to pathological fracture, dislocation, epidural hematoma, syringomyelia, platybasia, or

sarcomatoid transformation. Biochemical markers are useful in evaluating treatment effectiveness (8). In this case, the neurological deficits disappeared following surgical decompression, and post-surgical treatment with antipagetic drugs lowered alkaline phosphatase levels to normal.

Finally, urgent decompressive surgery is the treatment of choice in cases of spinal Paget's disease with a foot drop due to a pathological fracture, and medical antipagetic therapy should be given after decompression. We also emphasize that Paget's disease should be considered when a neurosurgeon encounters a foot drop.

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