



LUMBAR VERTEBRAL PAGET'S DISEASE TREATED WITH PERCUTANEOUS TRANSPEDICULAR SCREW FIXATION AND VERTEBROPLASTY

PERKÜTAN TRANSPEDİKÜLER VİDA TESPİTİ VE VERTEBROPLASTİ İLE TEDAVİ EDİLEN LOMBER VERTEBRA PAGET HASTALIĞI: OLGU SUNUMU

Ahmet KARAKAŞLI¹,
İsmail Safa SATOĞLU¹,
Ömer AKÇALI¹

¹Department of Orthopaedic Surgery, Dokuz Eylül University Faculty of Medicine, İzmir.

SUMMARY

Purpose: We describe a case with monostotic Paget's disease suffering from lumbar radiculopathy due to L4-5 instability and vertebral body collapse. Paget's disease is a chronic focal skeletal disorder that usually affects the pelvis and spine. Spinal cases are generally asymptomatic; in symptomatic cases, neurological dysfunctions are related to non-compressive vascular defects, hemorrhage, sarcomatous degeneration, spinal stenosis, or pathological fractures, primarily in the lumbar region.

Methods: In the present paper, a case of monostotic Paget's disease suffering from lumbar radiculopathy due to L4-5 instability and vertebral body collapse is described.

Results: The patient was treated with a hybrid surgical procedure combining percutaneous transpedicular screw fixation and vertebroplasty.

Conclusions: Paget's disease may lead to vertebral body collapse and foraminal narrowing, causing radiculopathy. Posterior spinal instrumentation with lordotic rods and vertebroplasty may reduce the pain and resolve the neurological deficit in patients with Paget's disease.

Key words: Paget's disease, vertebroplasty, lordotic rods

Level of evidence: Case report, Level IV

ÖZET

Amaç: Bu yazıda L4-L5 instabilitesi ve vertebra çökme kırığına bağlı radikulopati bulguları olan bir monostotik Paget Hastalığı olgusu tanımlanmıştır. Paget Hastalığı genellikle pelvis ve omurgayı etkileyen kronik, fokal bir iskelet bozukluğudur. Omurga vakaları genellikle asemptomatiktir; semptomatik vakalarda nörolojik bozukluklar çoğunlukla lomber bölgenin nonkompresif vasküler defektlerine, hemorajiye, sarkomatöz dejenerasyona, spinal darlığa ve patolojik kırıklara bağlıdır.

Metod: Bu makalede L4-L5 instabilitesi ve vertebra çökme kırığına bağlı radikulopati bulguları olan bir monostotik Pajet Hastalığı olgusu sunulmuştur.

Sonuç: Hasta cerrahi olarak perkütan transpediküler vida fksasyonu ve vertebroplasti ile tedavi edildi.

Çıkarımlar: Paget Hastalığı vertebra korpusunda çökmeye ve foraminal daralmaya yol açarak radikulopati sebebi olabilir. Lordotik rodlarla posterior spinal enstrümantasyon ve vertebroplasti ağrıyı azaltabilir ve nörolojik kayıpların düzelmesini sağlayabilir.

Anahtar Sözcükler: Paget Hastalığı, Vertebroplasti, Lordotik rod

Kanıt Düzeyi: Olgu sunumu, Düzey IV

Address: İsmail Safa Satoğlu,
Ortopedi ve Travmatoloji Anabilim
Dalı, Dokuz Eylül Üniversitesi Tıp
Fakültesi, İzmir.
Tel.: 0232 4123372
Gsm: 0532 6310087
E-mail: ismailsafo@yahoo.com
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INTRODUCTION:

Paget's disease of bone, or osteitis deformans, is a monostotic or polyostotic bone disorder, first described by Sir James Paget in 1877 and characterized by abnormal bone remodeling due to abnormal osteoclast function and morphology. Rapid bone resorption and deposition resulting in a mosaic pattern of lamellar bone, with extensive local vascularity and fibrous tissue in the bone marrow, is characteristic¹². The disease is polyostotic in 66% of cases and 35–50% show spinal involvement¹. When the spine is involved, the typical abnormal remodeling of the bone can lead to spinal stenosis with cord compression or radiculopathy. The incidence of spinal stenosis with cord compression in pagetic patients is 6%, and this can be caused by vertebral enlargement, intraspinal pagetic soft tissue overgrowth, ossification of epidural fat, epidural hematoma, and/or pathological fractures^{1,10,11}. Vertebral pathological fracture with spinal instability is one of the complications of Paget's disease and can lead to neurological compromise. Vertebroplasty is one of the options to control vertebral body collapse². If the patient has neurological compromise, decompressive procedures are advocated¹⁴. A pagetic vertebral motion segment is generally accepted as biomechanically stable, and instrumentation is rarely necessary¹³. However, vertebral body collapse resulting from pagetic fracture may require stabilization.

Here, we describe a case with monostotic Paget's disease (PD) suffering from lumbar radiculopathy due to L4–5 instability and vertebral body collapse. He was treated with a hybrid surgical procedure combining posterior percutaneous pedicular screw fixation and vertebroplasty. The neurological status of the patient improved without requiring any further procedures for decompression surgery.

CASE REPORT:

A 46-year-old male patient was admitted to our clinic with complaints of pain in the lower back and numbness for two years previously. Physical examination did not reveal any obvious neurological

deficit. Direct X-rays of the lumbosacral spine showed increased antero-posterior and transverse diameters, with expansion both in the vertebral body and the neural arch at the L4 vertebra, and osteosclerosis at the same level. The vertebra was enlarged with apposition of bone with respect to the cortical borders. Magnetic resonance imaging (MRI) revealed a diffuse medullary expansion with a rough trabecular pattern on the corpus and the posterior components of the L4 vertebra, causing relative spinal canal stenosis and dural sac compression with an intramedullary hyperintense signal on T2-weighted images at the L4 level (Figure-1).

At that level, the dura was compressed between the enlarged body and the anteriorly displaced L4 posterior arch. A bone scan (^{99m}Tc-bisphosphonate) showed increased uptake at the L4 vertebral body, confirming the pathological findings. Bone markers were checked, and the total alkaline phosphatase was 380 U/L and the calcium level was normal.

The differential diagnosis included sclerotic metastasis, lymphoma, osteosarcoma, giant cell tumor, and Paget's disease. Because of the clinical progression and the concomitant spinal instability, surgical management was proposed in the first instance.

This allowed spinal cord decompression and dorso-lumbar junction stabilization as well as pathological tissue sampling.

Histopathological examination showed irregular, disorganized bone lamellae with scattered, dissociated, plump pagetic cells. The presence of hypercellular bone trabecules in a woven pattern confirmed the diagnosis of Paget's disease (Figure-2).

Posterior pedicle screw fixation with distraction between the L3–5 levels and vertebroplasty at the L4 vertebra were planned (Figure-3). Polymethyl methacrylate was delivered using fluoroscopic guidance through both pedicles in the vertebral body of L4.



Figure-1. MRI showing rough trabecular pattern on the corpus and the posterior components of the L4 vertebra.

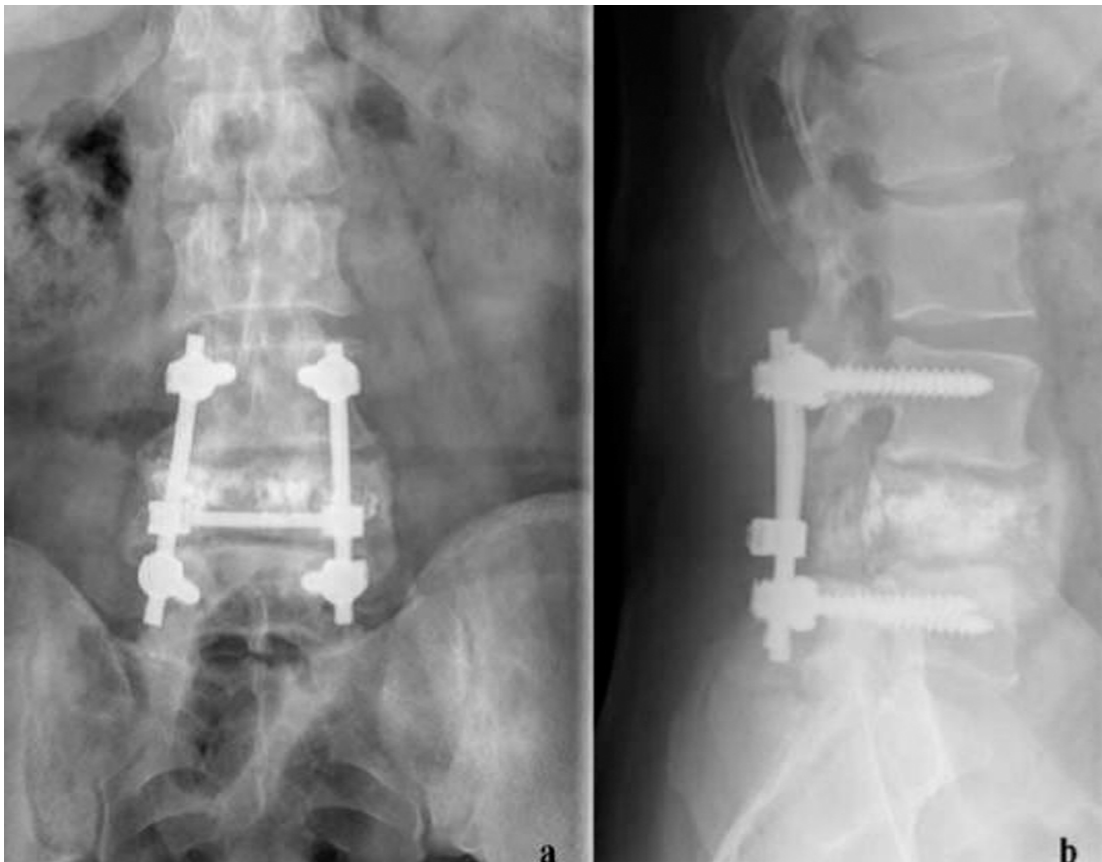


Figure-2. Posterior pedicle screw fixation with distraction between the L3-5 levels and vertebroplasty at the L4 vertebra.

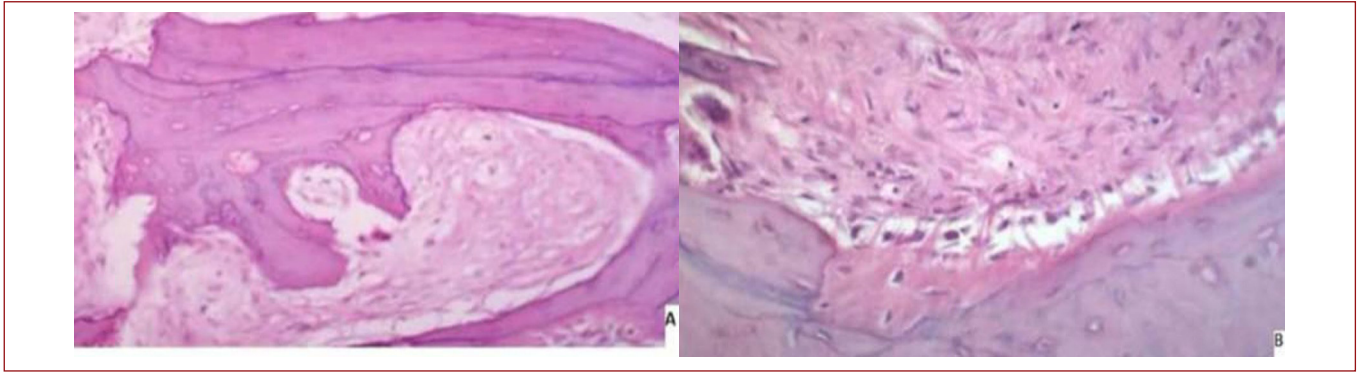


Figure-3. a) Osteoblastic and osteoclastic activities with irregular, disorganized bone lamellae (H&E, original magnification 100×). **b)** Surgical material revealing a woven bone pattern that confirms an active pagetic lesion.

Intraoperative and postoperative imaging revealed good distribution of the cement through the lytic areas of the vertebral body. Postoperatively, daily sodium etidronate treatment was started.

The postoperative course of the patient was uneventful, and he was discharged from the hospital one day after surgery. The treatment response was good, his symptoms quickly improved, and he had no neurological deficit eight months after surgery.

DISCUSSION:

Paget's disease is a chronic focal disorder of accelerated skeletal remodeling that can involve single or multiple bones and presents as enlarged or deformed bones at one or more skeletal sites. The underlying pathophysiological mechanisms of this disease are still unclear, but include genetic factors and paramyxovirus infection. The overall prevalence of Paget's disease is 3–3.7%. The disease is diagnosed over the age of 40 years in 90% of cases^{3,8}. The polyostotic form accounts for 66% of cases. The most commonly involved bones are the pelvis, vertebrae, skull, femur, and tibia³. In this case, the L4 vertebra was involved and the diagnosis was chronic progressive monostotic Paget's disease. Magnetic resonance imaging (MRI) scans showed a collapse at the L4 vertebra in the anterior and posterior portions, with a narrow focal stenosis of the spinal canal and compression of the medulla. Pagetic osteoclasts are abnormal, approximately five

times larger than normal and containing an average number of 100 nuclei per cell, as opposed to 3–4 nuclei per cell in normal adult osteoclasts, and have increased bone resorbing activity^{3,8,11}. However, the osteoblasts, though numerous, are not abnormal. Because bone resorption triggers bone formation, the rate of bone resorption is matched by a rapid rate of bone formation over time. Because of the dramatic increase in bone turnover, collagen fibers are deposited haphazardly during the process of bone formation, producing a woven bone, rather than a normal lamellar bone in which the collagen fibers are laid down in well-organized parallel bands¹¹. As a marker of osteoblast activity, the measurement of serum total alkaline phosphatase activity provides a general evaluation of bone turnover and disease activity in Paget's disease. Serum bone specific alkaline phosphatase activity is helpful in patients who also have liver disease or a plasma total alkaline phosphatase activity within the normal range^{3,11}. A rapid improvement of the clinical symptoms and a decrease of the alkaline phosphatase levels are also commonly seen after adequate antiresorptive therapy⁷. After the pelvis and long bones, the spine is the second most common site of involvement of Paget's disease. The lumbar region (58%) is the spinal region involved most often, followed by the thoracic (45%) and cervical (14%) regions^{9,11}. Pathological vertebral fracture with spinal instability is one of the complications of Paget's disease and can lead to neurological compromise.

Our case had an L4 pagetic collapsed vertebra and bilateral foraminal stenosis between the L4 and L5 levels. Spinal canal stenosis generally resolves with direct decompression with laminectomy and foraminectomy. In our case, we did not perform foraminectomy. Anterior support of the spinal column was achieved with percutaneous vertebroplasty. Vertebroplasty is a well-established technique that has been reported to provide significant relief to patients with painful vertebral compression fractures due to osteoporosis, multiple myeloma, hemangioma, or metastasis^{2,5,6}.

Posterior instrumentation with lordotic shaped rods may help to increase the sagittal diameter of the intervertebral foramina. Thus, neurological complaints caused by foraminal stenosis may resolve. This is thought to be a potential reason for the neurological improvement of patients. Here, we describe a case in which vertebroplasty combined with posterior pedicle screw fixation was useful in the treatment of vertebral collapse due to Paget's disease in a neurologically disabled patient. This hybrid surgical procedure combining posterior spinal instrumentation and vertebroplasty seems to be applicable to patients with or without pain and other neurological deterioration. Careful patient selection is crucial to achieve satisfactory results.

Severe comminuted vertebral body fractures necessitating anterior column reconstruction may still require combined surgical procedures.

In conclusion, Paget's disease may cause vertebral body collapse and foraminal narrowing leading to radiculopathy. Posterior spinal instrumentation with lordotic rods and vertebroplasty may reduce the pain and dissolve the neurological deficits in patients with Paget's disease. Screw-rod reduction before bone cement infusion provides a benefit in that the posterior longitudinal ligament can be propped open to enhance the strength of the posterior wall by letting the retropulsed bony fragments fall in its place. Short-segment fixation was performed in this case in order to reset the fracture, which might be the cause of active pain.

Therefore, for vertebral Paget's disease patients with posterior vertebral wall ruptures, screw-rod reduction before vertebroplasty is a better treatment option. Clinical experts have pointed out that vertebroplasty cannot achieve a satisfactory reductional effect on compressions of more than 80%, and so some physicians adopt vertebroplasty combined with postural reduction.

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