

MONOSTOTIC PAGET'S DISEASE IN LUMBAR VERTEBRAE: AN ATYPICAL LOCATION

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ABSTRACT

A 41-year-old white female patient with complaints of lumbar pain for more than three years, without irradiation, underwent several radiological examinations. Her condition was diagnosed

by means of biopsy, as monostotic Paget's disease in the third lumbar vertebra. This is an uncommon location.

Keywords – Paget's disease; Monostotic fibrous dysplasia; Spine

INTRODUCTION

Paget's disease was first described by Sir James Paget in 1877, under the name osteitis deformans. It consists of a bone remodeling disorder. There are etiological hypotheses involving viruses, mutations and/or genetic heredity. It is more common among white individuals, in the age group over 60 years and among men, in the proportions of 3:2. In most cases, it can be diagnosed using simple radiography. The alkaline phosphatase concentration is an important marker for controlling systemic forms. The current treatment consists of using bisphosphonates and analgesics. Zoledronic acid has been used with a good response⁽¹⁻³⁾.

The monostotic form is rare and is most common in the long bones and pelvis. In the spine, it represents 10 to 15% of this form of presentation⁽⁴⁾.

The aim of this study was to report on the case of a patient with monostotic Paget's disease in the spine.

CLINICAL CASE

The patient was a 41-year-old white woman with a complaint of lumbar pain for more than three years. She underwent clinical treatment of the symptoms and

physiotherapy at that time. In the physical examination, she presented limitations regarding trunk flexion, pain upon local mobilization, rectification of lumbar lordosis, paravertebral contracture and normal muscle strength and sensitivity. She did not have any other signs or symptoms. There were no significant personal and/or family antecedents. Simple radiography showed a blastic lesion on the L3 vertebral body in the anteroposterior and profile views. Magnetic resonance imaging (MRI) (Figure 1) showed a lesion with a fracture, without compromising the medullary canal. Bone scintigraphy indicated a single lesion (Figure 2). The case was discussed with the Orthopedic Oncology group, and screening was applied because of a hypothesis of metastatic bone lesion. All the examinations were shown to be normal, including mammography, chest and abdominal tomography, laboratory tests and myelogram. It was decided to perform a biopsy guided by tomography, and this produced an inconclusive result. A transpedicular biopsy using a 4 mm trephine was then performed. The histopathological evaluation on the surgical specimen, under a conventional optical microscope, showed the presence of bone trabeculae of irregular shape and disorganized structure, with the formation of cement lines compatible with Paget's disease (Figure 3). The team

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Figure 1 – T1 and T2-weighted nuclear magnetic resonance images showing lesion in the third lumbar vertebra

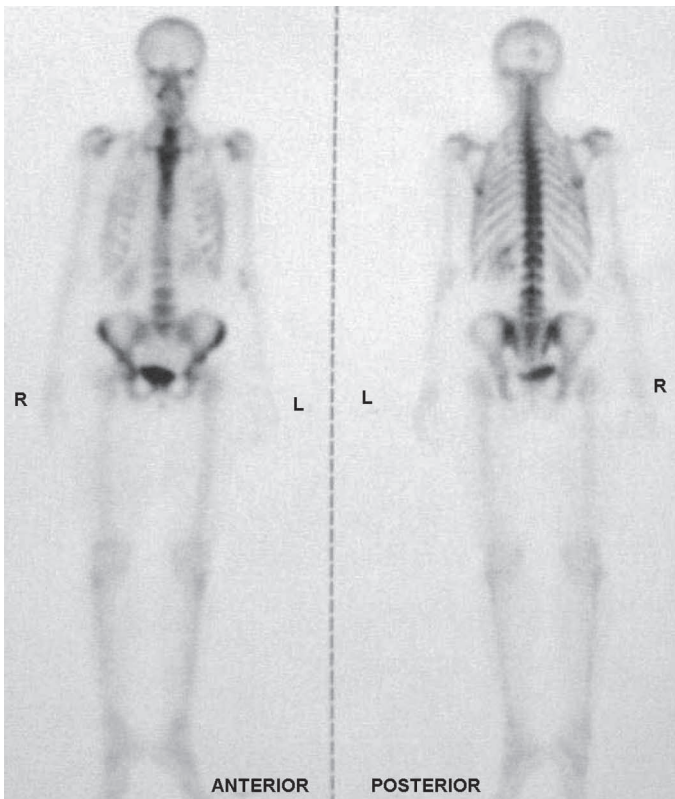


Figure 2 – Bone scintigraphy using technetium 99, with abnormally high uptake in L3

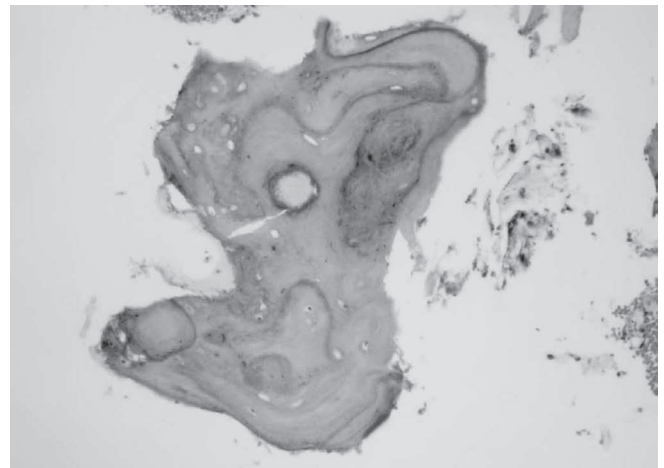


Figure 3 – Photomicrograph (HE; 100x) showing histological appearance of Paget's disease

decided to carry out internal stabilization using pedicle screws and nails because of fear of fractures, both of the pedicle and due to vertebral failure in the future, with the aim of avoiding new surgical procedures (Figure 4). The patient evolved well with the use of sodium alendronate, and with outpatient return visits.

DISCUSSION

Vukasinović *et al*⁽⁵⁾ reported the case of a patient with a hypothesis of disease in the parathyroids, for whom biopsy confirmed that it was a case of monos-

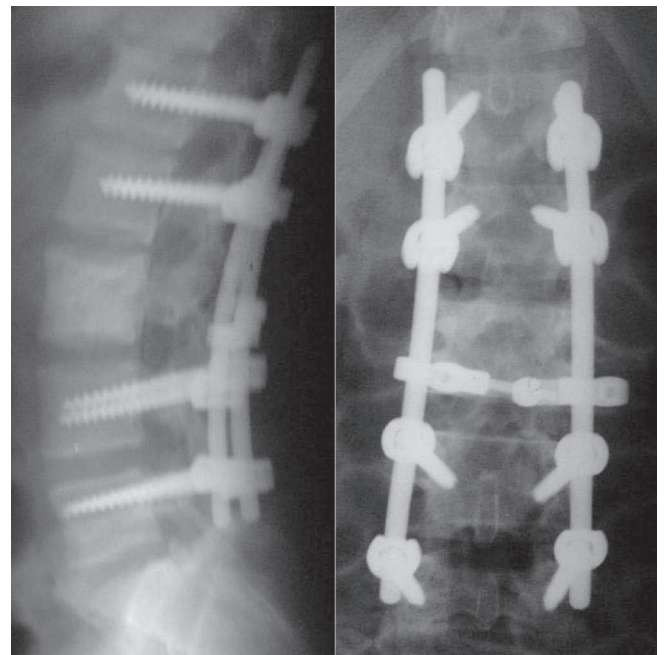


Figure 4 – Surgical treatment with pedicle screws and longitudinal nails

totic Paget's disease. In their paper, they recognized the benefits of early surgical stabilization, given that one of their patients who did not undergo an operation subsequently evolved to kyphosis. After receiving the diagnosis, our patient was given information about the natural history of the disease, its risk factors and its orthopedic management with corsets and surgery using screws or nails. Together with the medical team, our patient chose a surgical approach because of the greater stability possible, given that the pagetoid bone could in the future evolve to fracturing.

All patients with suspected Paget's disease should be evaluated by an oncological orthopedist, given that the hypothesis of neoplasia should thus be discarded. Reyes *et al*⁽⁶⁾ drew attention to the risks of diagnoses made from images suggestive of Paget's disease, and they reported the case of a patient with metastasis from hepatocellular carcinoma that simulated typical Paget vertebrae. There are other reports in the literature that confirm our opinion, in which the differential diagnosis between Paget's disease and bone neoplasia often becomes difficult⁽⁷⁾.

The vertebral lesions of Paget's disease may appear in different forms: lytic, blastic and/or mixed. Detailed studies need to be made, including biopsy whenever possible. The vertebral locations ranged from the axis

to the sacrum. Cases from outside of the standard age range are increasingly being found. Such cases may be diagnosed late, with neurological lesions already established⁽⁸⁻¹¹⁾.

Tomography-guided biopsy is the gold standard today for diagnosing vertebral lesions, but pathologists sometimes need large samples of material in order to reach a diagnostic conclusion⁽¹²⁾. In our case, we had to perform a biopsy using a 4 mm trephine, by means of the posterior transpedicular route, with an oncological orthopedist analyzing the macroscopic appearance of the sample during the operation.

In an interesting survey conducted in the United Kingdom among 1225 patients with Paget's disease, the most frequently found locations were the pelvis, lumbar spine, sacrum, femur, cranium and dorsal spine. It was found that 30% of the cases were of the monostotic form⁽¹³⁾. This percentage was much greater than found by most other authors. This finding raises questions relating to whether racial or genetic characteristics might be the causes.

Monostotic forms of Paget's disease should form part of the differential diagnosis for bone lesions, given that ever greater numbers of very atypical cases such as the case in the present report are appearing.

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