Monostotic Paget's Disease of the Lumbar Spine Mimicking Vertebral Metastasis: A Case Report

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Paget's disease of the bone is the second-most-common metabolic bone disease. However, it is rarely found in the Far East. Vertebral Paget's disease usually occurs at multiple vertebral levels, with less than 20% of vertebral Paget's disease being monostotic. Here, we present a rare case of monostotic vertebral Paget's disease, which we initially misdiagnosed as a spinal metastasis. A 34-year-old man was admitted with a one-month history of lower back pain. Initially, computed tomography of the lumbar spine showed an osteolytic change of the L4 and mild expansion of the L4 vertebral body. Subsequently, magnetic resonance imaging showed a highly homogeneously enhanced L4 vertebral body. We performed positron-emission tomography, bone scan, and tumor marker evaluation. However, we could not detect any likely primary origin of the spinal metastasis. Therefore, we carried out a needle bone biopsy under fluoroscopic guidance, and the bone specimen revealed Paget's disease of the bone.

Key Words: Paget's disease • Lumbar vertebra • Bone scintigraphy

INTRODUCTION

Paget's disease of the bone (PDB) is the second-most-common metabolic bone disease, surpassed only by osteoporosis. Among individuals of Anglo-Saxon origin older than 55, 3-3.7% suffer from PDB, but its prevalence is low in the Far East^{2,4,17}. Though PDB in the spine is not rare, monostotic vertebral Paget's disease (PD) definitively diagnosed through imaging studies or blood chemistry is quite rare. Here, we present a case of monostotic vertebral PD in the fourth lumbar vertebra that physicians initially misdiagnosed as a spinal metastasis.

CASE REPORT

A 34-year-old man was admitted to our clinic with a one-month history of lower back pain. His symptoms were continuous, dull pain. However, the patient did not present any

other motor, sensory, or sphincter abnormalities. A simple X-ray of the lumbosacral region showed a mildly radiopaque, sclerotic-margined L4 vertebra and a radiolucent change in the L4 vertebral body's inner portion as compared to other lumbar vertebrae. However, by roentgenographic analysis the L4's gross appearance showed no significant change indicative of any bone pathology. After these tests, the patient underwent lumbar spine computed tomography (CT) and this scan showed osteolytic changes in the L4 vertebral body's inner trabecular portion and mild expansion of the L4 vertebra (Fig. 1). Subsequently, the patient underwent gadolinium-enhanced magnetic resonance imaging (MRI) of the lumbar spine. The MRI revealed a highly enhanced L4 vertebral body, including posterior elements such as the pedicle, lamina and spinous process, suggestive of a hypervascular bone lesion with destructive changes of the vertebral body (Fig. 2).

Initially, we thought the lesion was a spinal metastasis. Therefore, the patient underwent on a positron-emission tomography (PET) scan, bone scintigraphy and serum tumor marker evaluation, to detect the primary origin site. However, the PET scan revealed a hypermetabolic lesion on L4 only, and bone scintigraphy showed non-specific findings except for the accumulation of the radionuclide in the L4 vertebra (Fig. 3). All serum chemistries, including tumor markers, were within normal ranges. Therefore, we performed a needle bone biopsy of the L4 vertebral body to obtain a bone specimen,

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Fig. 1. Coronal computed tomography scan, demonstrating expansion of the transverse diameter and the osteolytic change of the L4 vertebral body.

Fig. 2. Gadolinium-enhanced magnetic resonance imaging shows a highly enhanced L4 vertebra, including its posterior element, on sagittal section (A) and with destructive changes in the L4 vertebral body on the axial image (B).

under fluoroscopic guidance. We transferred the specimen to a pathologist, and it revealed the patient's PDB (Fig. 4). Hence we decided to treat the patient with oral bisphosphonate. In two months of treatment, his back pain had improved gradually from 8 to 2 scores on visual analogue scale.

DISCUSSION

First described by Sir James Paget, PDB is characterized by excessive and abnormal remodeling of bone due to enhanced resorption by giant osteoclasts, followed by abundant new bone

Fig. 3. The PET scan reveals hypermetabolism in the L4 vertebra (A) and the bone scan shows the radionuclide accumulation in the L4 vertebra alone (B).

Fig. 4. Sections reveal irregularly thickened bony trabeculae with prominent osteoblastic (opened arrow) and osteoclastic activity (closed arrow). (H&E, $\times 100$).

formation^{13,15}. PDB is the most common metabolic bone disease after osteoporosis, but its incidence varies according to populations' genetic backgrounds and environmental factors¹¹. PDB appears most often in populations of Anglo-Saxon origin and rarely in the middle and Far East, Africa, or Scandinavia^{1,2,4,11,17}.

The spine is the second-most-commonly affected site, after the pelvis and 66% of PDB cases have polyostotic involvement^{2,8,14}. Therefore, although the incidence of pagetic involvement of the vertebra is not low, monostotic vertebral PD is rarely reported, especially in the Far East, such as in our case.

Usually, patients with vertebral PD are asymptomatic, and the disease is detected through incidental findings of an elevated alkaline phosphatase (ALP) level, radiologic features, and bone scintigraphy⁶. Researchers have considered total serum ALP level to be a useful biomarker for PDB. Historically,

elevated total ALP activity has been of value in the diagnosis of new PDB patients. However, monostotic PDB may correlate with total ALP levels within the reference interval, introducing difficulties in both the diagnosis and follow-up management of such patients⁹. In fact, our patient's total ALP level was within normal values (220 U/L, normal range 90-250 U/L). In the disease's later phase, the vertebral body shows a typical "picture frame" appearance radiographically, due to the combination of trabecular bone hypertrophy and thickening at the end-plate, with apposition/absorption on the periosteal/endosteal surfaces at the anterior and posterior vertebral borders^{2,3,17}. However, in PD vertebral body involvement's early radiographic appearance is a thickening and hypertrophy of the trabecular bone, parallel to the end plate, which can appear similar to a thickened cortex^{2,3,13}. Radionuclide bone scans help to establish the diagnosis, and the physician should perform such scans in all PD patients, to determine the disease's distribution¹⁷. Two distinct pathognomic vertebral bone scan images are highly specific for PD. One is the "clover" image, affecting both vertebral pedicles and the spinous process and the other is the "heart" image, affecting the vertebral body and the spinous process^{7,10,12}. Previously reported specificity of these images for vertebral Paget's disease diagnosis is 100%. However, not only do these cases typically show pathognomic images only rarely, but also these pattern types appear in some patients with vertebral metastasis^{7,10}. In our patient, all portions of the L4 vertebra accumulated radionuclide on the bone scan, so we did not have any pathognomic findings that revealed specific disease entities.

PD is primarily a disorder of the bone, not of the bone marrow. However, secondary bone marrow changes can occur. In latter stages of vertebral Paget's disease, fatty transformations of bone marrow appear on MRIs⁵. In osteolysis of pagetic vertebrae, a fat signal within the lesion is a useful clinical determinant for distinguishing between vertebral Paget's disease and osteolytic metastasis^{2,16}. However, as early Paget's disease is not always distinguishable from a metastatic tumor, physicians can biopsy the lesion.

Thus, the definitive diagnosis of vertebral monostotic PD through blood chemistry, imaging studies, or bone scans is quite difficult due to its rarity and heterogenous presentation on imaging and scintigraphic findings.

The spine is the second most commonly affected site in PDB, predisposing patients to low back pain and spinal stenosis and pagetic arthropathy is a major contributing factor to both back pain and spinal stenosis. Back pain in vertebral PD may also be attributed to blood engorgement of the vertebral body caused by vascular and disorganized, hyperactive remodeling process⁴.

Bisphosphonates are the mainstay of the treatment of

PDB^{4,8}. Randomized trials indicate that antiresorptive treatment reduces associated pain and the levels of alkaline phosphatase, a useful marker of disease extent and activity. Also treatment of pagetic spinal stenosis symptoms should start with medical antipagetic therapy^{4,8,13-15}. Calcitonin, mithramycin, sodium etidronate and clodronate have been reported to either improve or to completely reverse the clinical symptoms of spinal stenosis^{4,8,14}. However, relapse or aggravation of spinal stenosis symptomatology after medical antipagetic treatment is not uncommon. So in these case, decompression of spinal stenosis should be implemented promptly after failure of antipagetic therapy^{4,14}.

CONCLUSION

We present a rare case of monostotic vertebral PD initially diagnosed as vertebral metastasis. In treatment of monostotic vertebral lesions, the physician should bear in mind the possibility of pagetic involvement in vertebrae for successful diagnosis and proper management, even though PD's incidence is low, particularly in the Far East.

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