

case report

Radiographic, computed tomographic and magnetic resonance imaging appearances of primary V-cutting zone of resorption of lumbar vertebral body in Paget's disease

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Background. Paget's disease of bone typically demonstrates three evolutionary phases with the characteristic radiographic findings. The incipient stage is manifested by an advancing lytic zone of resorption. Unlike the skull and the long bones the primary advancing zone of rarefaction has not been clearly demonstrated within the spine and pelvis.

Case report. A 62-year-old man with histologically proved polyostotic Paget's disease was admitted to the hospital due to the exacerbation of pain at the thoracolumbar junction with deterioration during the night. Due to slight elevation of acid phosphatase and clinical signs indicating prostatic hypertrophy the possibility of metastatic prostatic carcinoma has been raised. A bone scintigraphy with technetium phosphonate showed the expected marked increased uptake affecting numerous bones with the typical radiographic signs of Paget's disease. Besides these findings, there was also moderate focal accumulation within the right dorsal half of the vertebral body of L1. Conventional tomography, computed tomography (CT) and magnetic resonance imaging (MRI) demonstrated the primary cutting zone of resorption affecting the same region.

Conclusions. We present a unique case of Paget's disease with primary V-cutting zone within the undeformed vertebral body of L1 and spontaneous progression to the midphase of the disease on a three year follow-up.

Key words: osteitis deformans; spinal diseases - radiography; tomography, x-ray computed; magnetic resonance imaging

Introduction

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Paget's disease of bone typically demonstrates three evolutionary phases with the characteristic radiographic findings. The initial lytic phase is followed by the midphase with a mixture of lytic and sclerotic changes leading finally to the late sclerotic phase of the disease. The incipient stage is manifested

by an advancing lytic zone of resorption. Unlike the skull and the long bones the primary advancing zone of rarefaction has not been clearly demonstrated within the spine and pelvis.¹ It has been stressed that the most of vertebral lesions reported in literature as the primary lytic phase of the disease actually represent the secondary bone resorption following the compression of the vertebral bodies.²

In this case report, we describe radiographic, computed tomographic (CT) and magnetic resonance imaging (MRI) findings of primary cutting zone of the resorption within the undeformed vertebral body of L1. A three-year follow-up revealed a progressive transition to the midphase of Paget's disease.

Case report

A 62-year-old man with histologically proved polyostotic Paget's disease was admitted to the hospital due to the exacerbation of pain at the thoracolumbar junction with deterioration during the night. The physical examination of the thoracolumbar spine demonstrated the limited mobility without neurological impairment. Pertinent laboratory findings included extremely elevated levels of serum alkaline phosphatase and urine hydroxyproline. Due to slight elevation of acid phosphatase and clinical signs indicating prostatic hyperthrophy the possibility of metastatic prostatic carcinoma has been raised. A bone scintigraphy with technetium phosphonate showed the expected marked increased uptake affecting the right frontal area, the vertebrae C3, T1, T5, L2, L3, L4, L5, the left scapula, the pelvic bones and the proximal part of the left femur. Besides these sites, there was also moderate focal accumulation within the right dorsal half of the vertebral body of L1. The plain films of the affected areas demonstrated features typical for the midphase of polyostotic Paget's disease with the exception of the L1

vertebra. A careful analysis of the conventional tomography of L1 revealed the discrete triangular osteolytic area affecting the right dorsal part of the vertebral body localized just below the lower end plate and the dorsal cortex (Figure 1a). The osteolytic area correlated well with the region of moderately focally increased uptake on the bone scintigraphy. MRI of L1 showed the triangular low signal intensity area on T1-weighted spin-echo images, which was of high signal intensity on T2-weighted images and demonstrated the intense Gd-DTPA contrast enhancement on T1-weighted postcontrast images (Figure 1b). Since the clinical, scintigraphic and radiologic findings were in favour of the eventual metastatic disease secondary to prostatic carcinoma CT guided bone biopsy was pro-

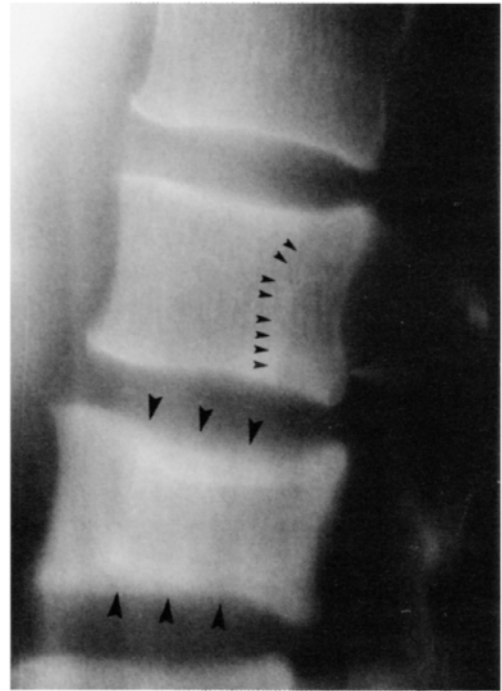


Figure 1a. Lateral conventional tomography of undeformed L1 vertebral body shows discrete triangular primary cutting cone zone of resorption localised below the lower endplate and the dorsal cortex (small arrow heads). Note also »picture-frame» appearance of the vertebral body L2 (big arrow heads), typical for the midphase of Paget's disease.

posed. The CT examination of L1 vertebra showed an oval purely lytic lesion below the dorsal cortex without any enlargement of bone contours (Figure 1c). The patient ultimately refused the bone biopsy and was discharged from the hospital. He was lost from our control for one and a half year. During that time the patient was treated intermittently with calcitonin and diphosphonates but laboratory examinations demonstrated only slightly decreased levels of serum alkaline phosphatase and of urine hydroxyproline. The repeated CT examination of L1 vertebra (Figure 2a) showed that the lesion progressed centrifugally for approximately 6-7 mm. The lesion, which at the first examination was purely lytic, on the control examination demonstrated mixed lytic/sclerotic features. The advancing edge of the lesion re-

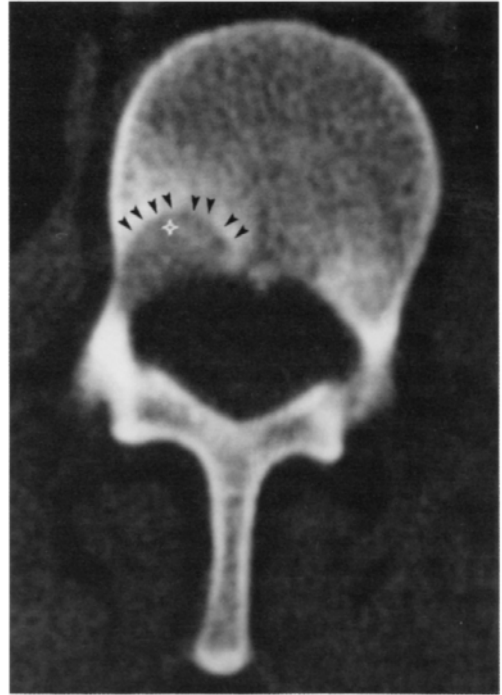


Figure 1c. Initial CT scan of L1 reveals an oval purely osteolytic lesion without evident bone enlargement (small arrow heads).



Figure 1b. Sagittal T1-weighted (500/15) spin-echo Gd-DTPA postcontrast image demonstrates marked contrast enhancement within the primary lytic zone of L1 (small arrow heads) and the central part of L2 vertebral body (big arrow head)

mained osteolytic and it looked as it was followed by osteosclerosis. In comparison with the initial examination the most interesting new finding was some enlargement of the bone contour seen along the posterior border of the vertebral body of L1 (Figure 2b). The repeated bone scintigraphy was similar to the first examination and revealed no additional focal accumulation, which could be expected in case of eventual metastatic disease. On a three year follow-up radiography, CT and MRI demonstrated the further progression of the lesion with a mixture of lytic/sclerotic findings typical for the midphase of Paget's disease. The slight compression of the lower end plate of L1 was evident reflecting the weakening of the pagetic bone (Figure 3).

Discussion

The primary lytic phase of Paget's disease of bone is characterised by an advancing zone of bone rarefaction reflecting the intense osteoclastic activity. The average rate of progression in the untreated patients has been estimated at about 1 cm/year (1mm /month). The focal bone resorption is quickly followed by a marked osteoblastic repair which tends to compensate the loss of the bone.³ This focal bone balance between resorption and formation may demonstrate considerable variations.² Therefore, the sclerotic phase may be quickly changed into the mixed or the lytic appearances of the disease. The advancing zone of rarefaction can be easily revealed radiographically within the skull as *osteoporosis circumscripta* or within the long bones as V-shaped cutting cone. However, within the tra-

becular bones like pelvis and spine, the primary lytic zone has not been clearly demonstrated.¹ The first radiologic evidences in these areas are typically sclerotic features of the disease.³ According to Maldague and Malghem² most of the vertebral lesions, reported in literature as demonstrating the cutting cone of primary lytic phase, actually followed the final sclerotic phase of the disease. The secondary resorption within the sclerotic pagetic bone usually occurs due to some provoking factors, most commonly the compression of the vertebrae, and has been never observed at the level of undeformed vertebral bodies. In this respect our case is unique. An osteolytic zone has been clearly demonstrated by the conventional tomography and CT examination within the undeformed vertebral body of L1 (Figures 1a, 1c). Furthermore the primary zone of rarefaction affected only a part of the vertebral body of L1. In other re-

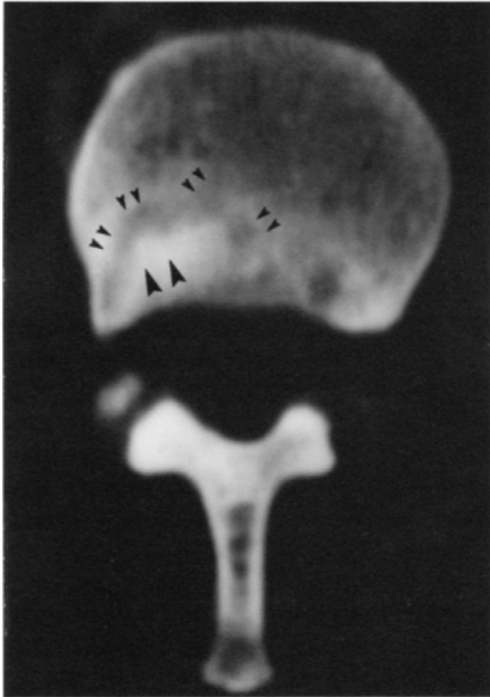


Figure 2a. Follow-up CT examination one and a half year later shows centrifugal enlargement of the lesion. The advancing lytic cutting cone (small arrow heads) is followed by osteosclerosis (big arrow heads).

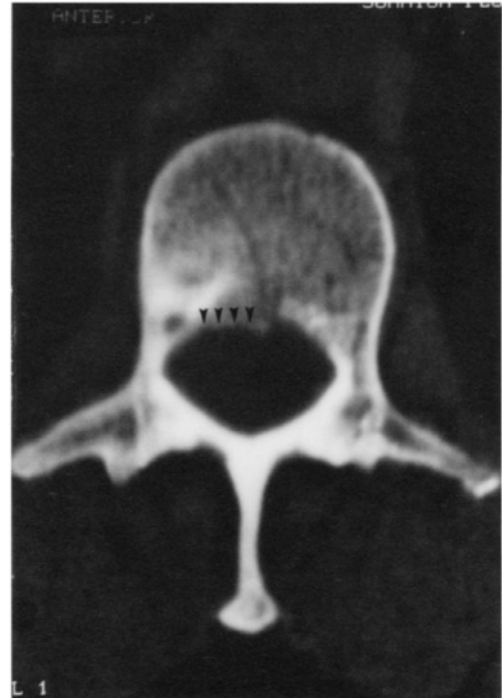


Figure 2b. CT scan also demonstrates an enlargement of bone contour at the dorsal aspect of the vertebral body of L1 (small arrow heads)

ported cases not only the compression of the vertebral bodies was seen, but the rarefaction was more diffusely distributed.^{2,4,5} There are no reasons to believe that in this particular case the zone of rarefaction appeared secondarily within the preexisting pagetic bone. At the initial examination the lesion was purely osteolytic and there was no enlargement of the vertebral body which would be present in a case of previous sclerotic phase of the disease. The scintigraphic finding of only moderately increased uptake of technetium phosphonate within the primary resorption zone seen in our patient has been reported in literature at the initial phase of Paget's disease.² MRI appearances of high signal intensity on T2-weighted images and

marked contrast enhancement on T1-weighted postcontrast images (Figure 1b) within the primary lytic zone of L1 and the central part of vertebral body of L2 represented oedema and hyperaemia consistent with the replacement of fat and haematopoietic elements of normal bone marrow with hypervascular fibroblastic tissue characteristic of Paget's disease.¹ The radiologic features on a follow-up examination were also in favour of the existence of primary lytic zone within the vertebral body of L1. Two of the four cardinal radiographic appearances of Paget's disease were shown, the enlargement of bone contours and an advancing resorption zone. A typical bone enlargement not seen at the initial examination appeared only after one and a half year reflecting activation of the subperiosteal cortex (Figure 2a). At that time there was also the evident progression of primary cutting cone for about 6-7 mm. The primary zone of resorption was quickly followed by secondary osteosclerosis, which may reflect a higher remodelling rate of the trabecular bone.² The rapid process of sclerosis could be eventually attributed to antiosteoclastic therapy. However, the influence of treatment was probably limited since the levels of serum alkaline phosphatase and urine hydroxyproline were not significantly decreased after one year and a half.

Without bone biopsy the definite diagnosis of Paget's disease still remains hypothetical. On the other hand the eventual bone biopsy at the beginning of the lesion of vertebral body of L1 could substantially change the natural evolution of the disease. Anyhow, the diagnosis of metastatic disease after a three year follow-up was not realistic. Relevant clinical, laboratory and scintigraphic findings were all negative. Besides, there were no radiologic signs of local cortical destruction and extension of the osteolytic lesion of L1 into the surrounding soft tissues which would be expected in the metastatic disease. The lesion has progressed within the periosteal envelope

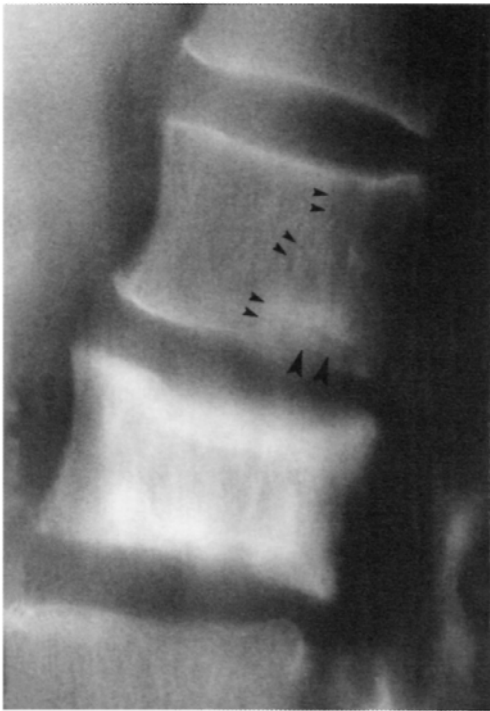


Figure 3. A three year follow-up lateral tomography reveals mixed lytic/sclerotic features typical for the mid-phase of Paget's disease. The lesion has progressed and affects nearly half of the vertebral body L1 (small arrow heads). Note initial concavity of the lower end plate in the region of weakened pagetic bone (big arrow heads).

of bone, findings characteristic of a midphase of Paget's disease. Finally, the radiological changes of the vertebral body of L1 on the follow-up examinations were comparable to the neighbouring vertebrae with all typical signs of a midphase of Paget's disease.

In summary, we presented a unique case of Paget's disease with primary V-cutting cone within the undeformed vertebral body of L1 and spontaneous progression to the midphase of the disease on a three year follow-up. Pertinent radiographic, CT and MRI findings were described.

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