

Unusual presentation of lumbar chordoma on bone scintigraphy in a young patient

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ABSTRACT

Chordoma is a rare bone cancer which arises from undifferentiated notochordal remnants in the axial skeleton. It generally has slow-growing and locally aggressive behavior. This tumor is usually diagnosed by CT and MRI modalities and the role of SPECT/CT is still debated. It shows reduced or normal uptake of radioisotope on bone scanning and increased tracer uptake is infrequently reported. Here we present a 33-year-old man with complaint of low back pain and numbness of his right leg. The whole body bone scan showed relatively uniform radiotracer activity throughout the skeleton. A focal increased uptake in the second lumbar vertebra was noted on SPECT/CT images. SPECT/CT also demonstrated multiple lytic lesions in lumbar vertebrae. The lesions were proven to be chordoma on biopsy. Lumbar chordoma could be one of the differential diagnoses for lytic lesions of the vertebrae which show absent or minimal tracer uptake on bone scintigraphy and SPECT/CT imaging. Our case was unusual as the patient was very young for chordoma diagnosis and bone scan showed increased uptake adjacent to the involved vertebral lesion detected by SPECT/CT.

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Introduction

Chordoma is a rare bone cancer with incidence of approximately 0.08 per 100 000, with predominance in men. Peak incidence has been reported to occur between 50-60 years of age (1, 2). No racial predilection has been found and the incidence of chordoma in men is 2-fold greater than in women (3, 4). The incidence of chordoma is very low in patients younger than 40 years, and children and adolescents are rarely affected. This tumor accounts for 1-4% of all malignant bone tumors (2, 5). Chordomas are thought to arise from undifferentiated notochordal remnants in the axial skeleton (6).

The clinical presentations vary and depend on location. Skull-base chordomas often present with cranial-nerve palsies. Some other manifestations are endocrinopathy, epistaxis and intracranial haemorrhage (7, 8). Chordomas of the mobile spine (cervical, thoracic and lumbar spine) and sacrum may present with localized

deep pain or neurologic symptoms with radiculopathy and/or bladder-bowel dysfunction (9). Historically, it was presumed that chordomas are presented more often in the sacrum and the skull base; however, recent evidence suggests almost equal distribution in the skull base (32%), mobile spine (32.8%), and sacrum (29.2%) (10).

Computed tomography (CT) and magnetic resonance imaging (MRI) have complementary role in diagnosis for chordoma (11) and these tumors generally show reduced or normal uptake of radioisotope on bone scanning (12).

Case report

We report a 33-year-old man with complaint of low back pain and numbness of the right leg since one month ago. He had no history of previous disease or drug consumption and his pain was not associated with trauma or fever. The patient was referred to our center for bone scintigraphy with presumed diagnoses of primary bone

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tumor, metastasis or osteodiscitis. The findings of the patient's MRI, which was done 1 week earlier, suggested destructive lesions in 1st, 2nd and 3rd lumbar vertebrae with involvement of the bodies and pedicles. These lesions showed high

signal in T2 and low signal in T1w sequences. No evidence of disk herniation, central canal or neural foraminal narrowing was reported (Figure 1).

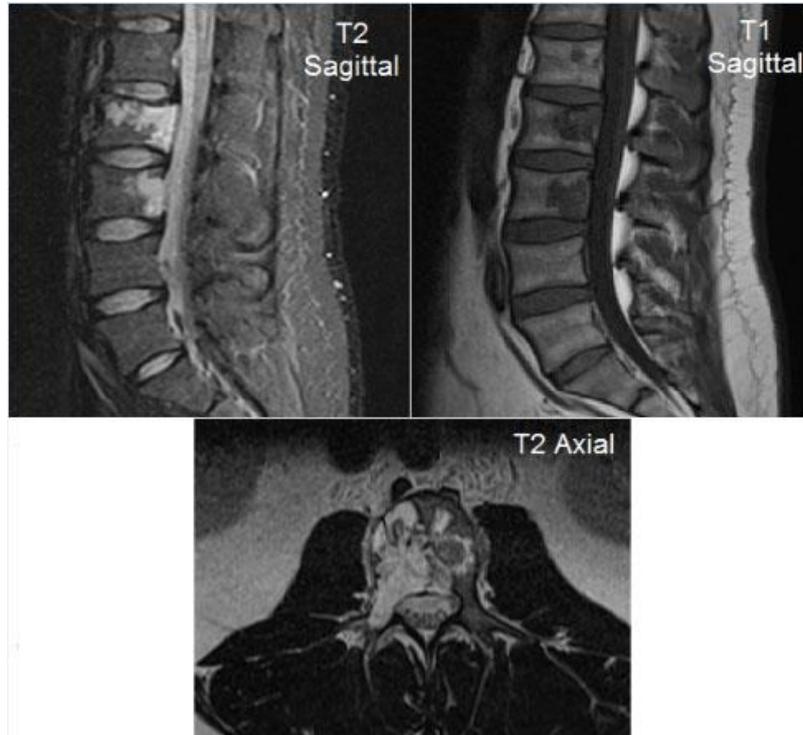


Figure 1. The lesions of the lumbar vertebrae showed high signal in T2 and low signal in T1w sequences. No evidence of disk herniation, central canal or neural foraminal narrowing was noticed

Three hours after intravenous injection of 740 MBq of Tc-99m methyl diphosphonate (MDP), whole body bone scan was performed using a dual-head gamma camera (GE) equipped with low-energy and high-resolution parallel-hole collimator (13 cm/min table speed, matrix size of 256×1024 and 140 keV energy window with 10% width) (Figure 2). SPECT images (128×128 matrix using 64 projections in a non-circular orbit with 20 seconds per step) were also performed and reconstructed by an iterative method (OSEM, number of iterations 8 subsets 4). The CT part of the SPECT/CT was done for anatomical correlation and attenuation correction (spatial resolution 3mm, 120 kV and 60-80 mAs) (Figure 3).



Figure 2. The whole body scan showed relatively uniform radiotracer activity throughout the skeleton, except for a faint focal uptake seen on the posterior view of the second lumbar vertebra (arrow)

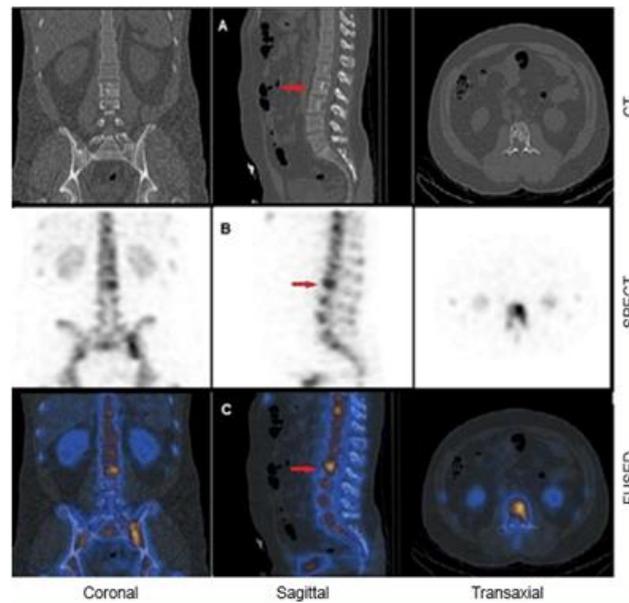


Figure 3. SPECT/CT imaging of the lumbo-pelvic region revealed multiple lytic lesions in 1st, 2nd and 3rd lumbar vertebrae. Increased uptake was noted in body of L2 adjacent to the lesion. Arrows show the lytic lesions on CT slices (A), increased uptake in L2 of SPECT (B) and overlaid SPECT/CT images (C)

The whole body scan showed relatively uniform radiotracer activity throughout the skeleton, except for a faint focal uptake seen on the posterior view of the second lumbar vertebra (Figure 2, arrow). SPECT/CT imaging of the lumbo-pelvic region revealed multiple lytic lesions in 1st, 2nd and 3rd lumbar vertebrae and a zone of increased uptake were noted in body of L2; which were confined to the sclerotic part of the lesion on CT component (Figure 3). Considering the tumoral involvement of the lumbar vertebrae and no history of previous malignancy, biopsy was done and the pathology was compatible with chordoma.

Discussion

Chordoma is known as a rare bone tumor; which is generally slow-growing and locally aggressive (10). However, the risk of metastasis is about 5% and occurs mostly to lung (9). Unfortunately, the non-specific nature and insidious onset of pain often delays the diagnosis until late in the disease course (9, 13). Surgical resection remains the primary and the main part of treatment (14) and radiation therapy as primary or adjuvant treatment is debated (15). The local control and tumor size at diagnosis, effect on surgical outcomes (16). Local control of the tumor is a major challenge in this disease, with a rate of more than 50% local relapses after complete surgery (17). This tumor is usually diagnosed by CT and MRI modalities, which are crucial imaging in diagnosis for chordoma (11);

however biopsy procedure and histological confirmation are necessary for accurate diagnosis (18).

Calcification and bony expansion are some diagnostic features of chordoma which show isointense or hypointense on T1-weighted MRI images and hyperintense on T2-weighted, and will enhance with gadolinium (10). It is very important to remember that chordomas generally show reduced or normal uptake of radioisotope on bone scanning and increased tracer uptake is infrequently reported (12, 19). Therefore, the diagnosis of this tumor on bone scintigraphy can be so challenging.

The role of molecular imaging in evaluation of chordoma is rarely explored. Some case reports show moderate pathological uptake of fluorodeoxyglucose (¹⁸F-FDG) uptake ($SUV_{max}=4.5$) in the primary tumour of sacrococcygeal chordoma (11) and suggested that FDG-PET/CT in combination with other imaging modalities increases the sensitivity for the detection of recurrent and metastatic disease. PET/CT provides both anatomic and metabolic information and was useful in evaluating the metastatic disease, biopsy planning and also for differential diagnosis between metastatic disease due to chordoma and second primary tumor (20). Some studies suggest that bone scintigraphy can be useful in the evaluation of sacral tumors and a midline sacral tumor that shows absent tracer uptake is very likely to be a chordoma (12, 19). However, the role of bone

scan and SPECT/CT for diagnosis and management of chordoma is still debated (12).

The presented case implies that lumbar chordoma could be one of the differential diagnoses for lytic lesions of the vertebrae which show absent or minimal tracer uptake on bone scintigraphy and SPECT/CT imaging. Our case was unusual as the patient was very young for chordoma diagnosis and the tumor didn't show a purely lytic pattern and increased uptake was noted in the sclerotic part of the lesion involving one of the vertebrae. This difference in manifestation might be due to the young age of the patient or the stage of the disease; which needs further studies.

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