CASE REPORT

Lumbosacral osteosarcoma with dural spread, skip lesions and intravascular extension: A case report

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SUMMARY

Primary osteosarcoma of the spine is indeed rare and only several sporadic cases have been reported. It tends to occur in a slightly older age group than those with appendicular skeleton tumours. We present here an unusual case of aggressive lumbosacral osteosarcoma in a young teenager complicated by extensive dural spread, skip lesions and intravascular extension. Although a histopathological examination is mandatory to establish the diagnosis, this case emphasises the need of imaging to ascertain the full extent of disease spread especially in deciding the type of treatment to be instituted and to evaluate the response to the treatment.

INTRODUCTION

Primary osteosarcoma of the spine is indeed rare and only several sporadic cases have been reported.[1] It accounts for 3.6-14.5% of primary spinal tumours and 0.85-3.0% of all osteosarcomas.[2-3] It frequently occurs in a slightly older age group than those with appendicular skeleton tumours. We present a case of aggressive lumbosacral osteosarcoma in a young teenager, and describe the diagnostic and radiological features of this rare scenario. To the best of our knowledge, a vertebral osteosarcoma complicated by extensive dural spread, skip lesions and intravascular extension has not been reported in previous literature.

CASE REPORT

A 13-year-old Indian girl presented to a private centre with a main complaint of abnormal gait described as walking with the body deviated to the right side for two-month duration associated with loss of appetite and weight. She has also complained of tightness of her right thigh. She denied having low back pain or lower limb numbness. There was no history of fever or trauma. On physical examination, the gait was unsteady. There was tenderness felt at the lumbosacral junction on deep palpation, however no swelling or deformity was seen at this region. The power of the right lower limb was 4/5, although the sensation was intact. The straight leg raising test was 30º bilaterally. Biochemical tests showed increased alkaline phosphatase, otherwise normal values of calcium and phosphorus. D-dimer was negative. She was treated as having muscle spasm and was started on physiotherapy. Nonetheless, her condition did not improve. A month later, a magnetic resonance imaging (MRI) of the lumbosacral spine was performed revealing enhancing bony lesion with cortical destruction involving L5 vertebra and sacrum. There was soft tissue component seen occupying the spinal canal and neural foramina at the level of L4 to S3, with presacral muscles infiltration (Figure 1).

She was immediately referred to the University Malaya Medical Centre for further management. All her baseline investigations were reexamined and interestingly, her lumbosacral spine plain radiograph which was initially read to be unremarkable revealed permeative sclerotic bone pattern in the sacrum. A subsequent contrast enhanced computed tomography (CT) of the thorax, abdomen and pelvis was performed which showed sclerotic bony lesions involving L4 to S5 vertebrae with extensive sunburst periosteal reaction. There was infiltration of the adjacent pelvic floor muscles and associated intramural thrombi seen in the left internal and external iliac veins, both common iliac veins and distal inferior vena cava (Figure 2). There was no evidence of distant metastases. Based on these imaging findings, a provisional diagnosis of lumbosacral osteosarcoma with dural spread, skip lesions and tumour thrombi was given. An urgent bone biopsy was performed and the histopathological findings showed a cellular tumour, composed of mainly spindle shaped and occasional ovoid cells, producing osteoid with residual normal cortical bone seen. The tumour cells possessed large pleomorphic nuclei and prominent nucleoli. Therefore, the diagnosis of osteosarcoma (osteoblastic variant) was confirmed.

She was started on anticoagulation and inferior vena cava filter was inserted. She was eventually started on chemotherapy (combination of Doxorubicin, Cisplatin and high dose Methotrexate). Bone scintigraphy revealed no evidence of distant bone metastasis. After the first cycle of chemotherapy, she underwent a repeated MRI of the lumbosacral spine which showed disease progression. There was extensive local invasion to the pelvic bones with encasement of bilateral iliac vessels, involving bilateral psoas and iliacus muscles, and both sacroiliac joints. The chemotherapy was further continued and in between the treatment, CT scans were performed which showed increasing local invasion as well as new lung metastases. There were no significant changes seen on the recent MRI although with completion of her chemotherapy. Second line

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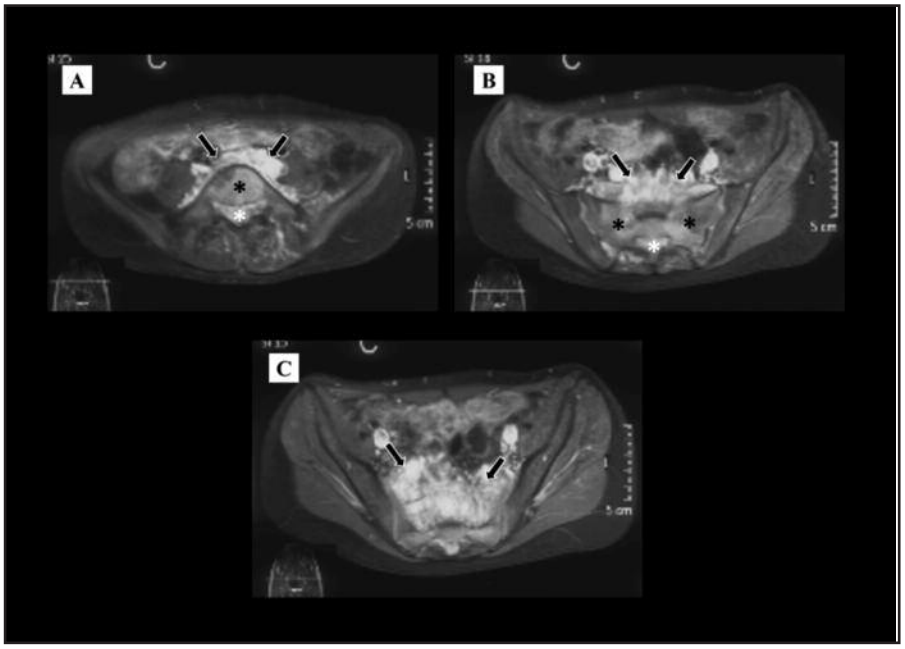

Fig. 1 : Axial T1-weighted post gadolinium sequence on MRI, showing enhancing bony lesion with cortical destruction involving L5 vertebra and sacrum (black asterisks) with soft tissue component seen occupying the spinal canal and neural foramina (white asterisks) on image A and B, as well as infiltration of the presacral muscles (black arrows).

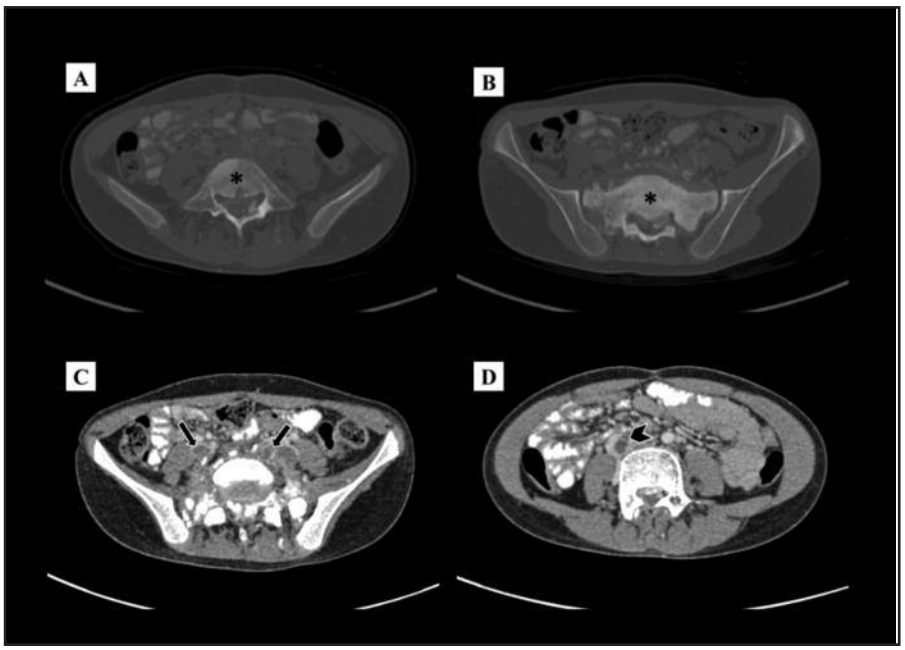

Fig. 2 : Axial contrast enhanced CT of thorax, abdomen and pelvis showing sclerotic bony lesions involving L4 to sacral vertebrae (black asterisks) with extensive sunburst periosteal reaction, on bone window (image A and B). On soft tissue window (image C and D), there was infiltration of the adjacent pelvic floor muscles and associated intramural thrombi seen in the left internal and external iliac veins, both common iliac veins (black arrows) and distal inferior vena cava (thick black arrowhead).

chemotherapy of Ifosfamide, Etoposide and Zoledronic acid was eventually started with further follow-up in our centre.

DISCUSSION

Osteosarcoma is a non-haematological primary malignancy of the bone and is mostly seen in the appendicular skeleton. Vertebral osteosarcoma which is indeed rare, appears to differ from osteosarcoma overall in its age distribution. It frequently affects those in their second to fourth decades of life, as opposed to our patient. The lumbosacral spine is the most common site followed by the thoracic and cervical spine respectively, with the vertebral body being the most commonly affected location.[3] The tumour may also extend into the pedicles and impinge on the spinal roots, or associated with an extraosseous mass as seen in our case. Skip metastases are synchronous smaller foci of tumour occurring in the same bone anatomically separated from the primary lesion, or as synchronous smaller foci of tumour on the opposing side of a joint. Extensive dural spread and tumour thromboembolism by osteosarcoma is extremely rare with only several sporadic cases have been reported.[4] In our case, there were presence of intraosseous and transarticular skip metastases involving several vertebrae with extensive multilevel dural spread and tumour thrombi involving the pelvic veins and distal inferior vena cava seen.

The clinical presentation may vary depending on the level of vertebral involvement with pain being the commonest presenting feature. Other features include neurological symptoms such as sensory deficits, motor abnormalities, and the presence of a palpable mass. Occasionally, these symptoms may be longstanding before the diagnosis is made, similarly as seen in our case.

Imaging modalities provide valuable information and are widely used to detect vertebral osteosarcoma.[3] Plain radiographs and CT images normally demonstrate a purely lytic, sclerotic or mixed lesion with compression of the involved vertebra.[2] CT allows identification of both matrix mineralisation and its invasion to the paravertebral and extradural soft tissues. MRI is used to evaluate the degree of tumour extension and to detect spinal canal involvement. The non-mineralised areas are non-specific; the lesions usually have low to intermediate signal intensity on T1- weighted and high signal intensity on T2-weighted images. Areas of bone formation may remain as areas of signal void on all pulse sequences. The plain radiograph in our case actually shows permeative sclerotic bone pattern in the sacrum. The disease was later confirmed by CT and MRI images whereby the tumour epicentre and origin was at the sacrum with evidence of skip lesions to the proximal lumbar vertebrae, local extension into the soft tissues and muscles anteriorly, and extensive intradural spread from L4 to S3.

It has been found that the prognosis for patients with osteosarcoma of the trunk is worse compared with the prognosis of patients with osteosarcoma of the extremities.[1-2] Treatment of vertebral osteosarcoma takes a multimodality approach as untreated case is universally fatal. Its course is marked by its aggressive local growth and rapid haematogenous spread. Lung metastases are the most common site of clinical significance as seen in our patient, whereby she was eventually diagnosed with it. Neoadjuvant chemotherapy is typically administered before resection of the primary tumour. However, the effectiveness of chemotherapy for osteosarcoma was found to be temporary and that regrowth is inevitable if the primary lesion was not resected.[5] Our patient was not keen for any surgical intervention. Hence, second line chemotherapy was given in view of poor response to first line chemotherapy and disease progression.

CONCLUSION

In view of the unusual presentation of this case, biopsy is definitely mandatory for the diagnosis. It also emphasises the need of imaging to ascertain the full extent of disease spread in a primary osteosarcoma of the spine especially in deciding the type of treatment to be instituted and to evaluate the response to the treatment. In conclusion, early detection and accurate diagnosis is indeed important to improve patients' prognosis and quality of life.

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