Solitary Spinal Extradural Plasmacytoma Causing Spinal Compression: A Case Report

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Citation

H Leung, S Chan, W Ng, H Law, K Li. *Solitary Spinal Extradural Plasmacytoma Causing Spinal Compression: A Case Report* . The Internet Journal of Orthopedic Surgery. 2024 Volume 32 Number 1.

DOI: 10.5580/IJOS.57077

Abstract

It is exceedingly rare for plasma cell neoplasm to present as solitary extradural plasmacytoma with only 10 case reports in literature. Here we report a rare case of solitary extradural plasmacytoma presented with bilateral lower limbs weakness.

Case Report

An 83-year-old man initially complained of subjective bilateral lower limbs weakness with unremarkable physical examination. He developed acute retention of urine and reduced lower limb power 3 days afterwards. MRI whole spine revealed a T7 to T9 extradural spinal tumour. Emergency operation of T7 to T9 laminectomy and spinal tumour excision was done. Pathology of the tumour was plasma cell neoplasm. He is now receiving palliative radiotherapy and thalidomide with multidisciplinary input from the medical and oncology colleagues.

Discussion

Solitary extradural plasmacytomas usually present as well-defined lesions located in the extradural dorsal spinal canal without bone erosion and bone marrow involvement. They have variable MRI features which can make pre-operative diagnosis difficult. Solitary extramedullary plasmacytoma is usually treated with surgery and radiotherapy, while the use of adjuvant chemotherapy remains controversial.

INTRODUCTION

Patients with plasma cell neoplasia can present with generalized disease in form of multiple myeloma, solitary bone plasmacytoma and even more rarely extramedullary plasmacytoma [1]. It is more common for plasma cell neoplasm to present in the spine as destructive osteolytic bone lesion causing spinal cord and nerve root compression [2] . On the other hand, extramedullary plasmacytomas arising from the spinal dura mater without surrounding bony destruction are defined as solitary extradural plasmacytoma. It is exceedingly rare and less than 10 cases could be found in English language. Here, we report a rare case of solitary plasmacytoma initially presented bilateral lower limbs weakness.

CASE REPORT

A 83-year-old men with history of common bile duct stones with endoscopic retrograde cholangiopancreatography done was admitted to the Department of Orthopedics and Traumatology for bilateral lower limb weakness. He had no back pain and no lower limb numbness. Initial physical examination upon admission revealed no tenderness at the lumbosacral spine. Lower limbs power was full in all myotomes. Reflexes were normal. Digital rectal examination revealed normal anal tone and grip, no saddle paresthesia was detected. X-ray of the lumbar spine showed no collapse with intact endplate and pedicles. Complete blood count, liver function test, renal function test and calcium level were unremarkable during admission.

3 days after admission, he was noted to have acute retention of urine with deteriorating lower limbs power bilaterally to 2-3/5 at the same day. Urgent MRI whole spine was done. According to the MRI report, an extramedullary contrast enhanced lesion which was isointense in T1-weghted image and slightly hyperintense in T2-weighted image was found from T7 to T9 and measured about 5.2cm in craniocaudal length, and 2.5 x 1.3cm on axial plane (Figure 1 and 2). The

spinal cord was markedly compressed by the tumor mass from T7/8 to T8/9 level and displaced to the left anterolateral aspect. T7 to T9 laminectomy with near total excision of spinal tumour was done by neurosurgeon. The tumour was found to be a yellowish lobulated epidural mass from T7 to T9 with good tissue plane from dura mostly over right side of spinal canal.

Figure 1

MRI Spine T1 imaging showing extramedullary mass lesion from T7 to T9 level with contrast enhancement

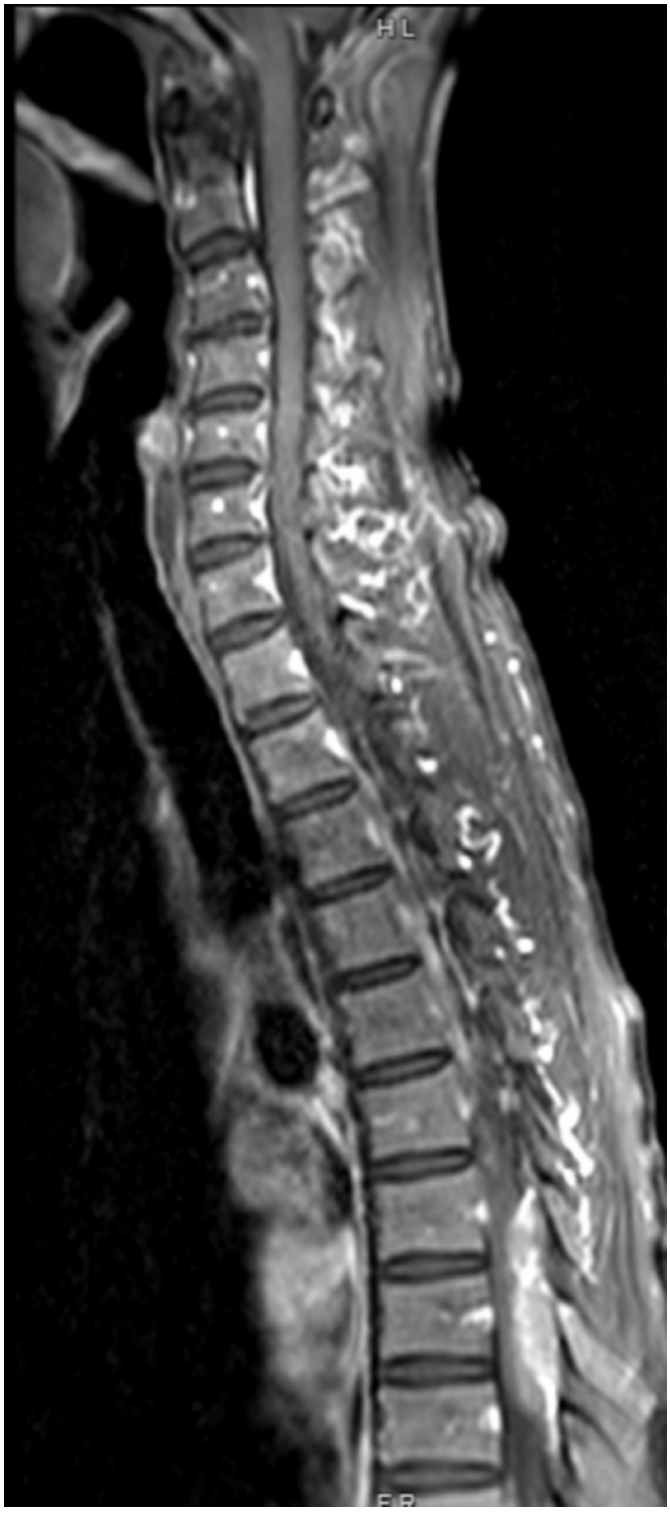


Figure 2

MRI Spine T1 imaging showing extramedullary mass lesion from T7 to T9 level with contrast enhancement

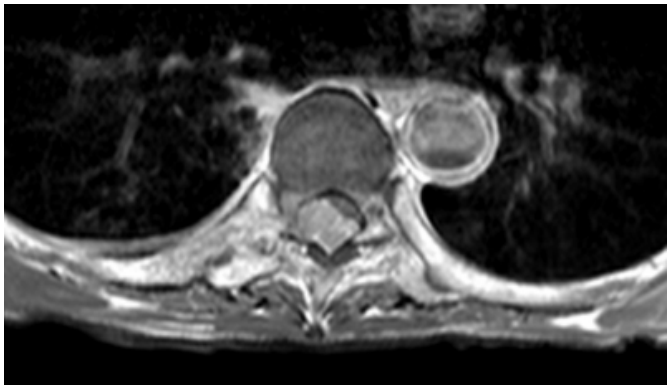


Figure 3

MRI Spine T2 showing a slightly hyperintense extramedullary lesion from T7 to T9 with compression on spinal cord.

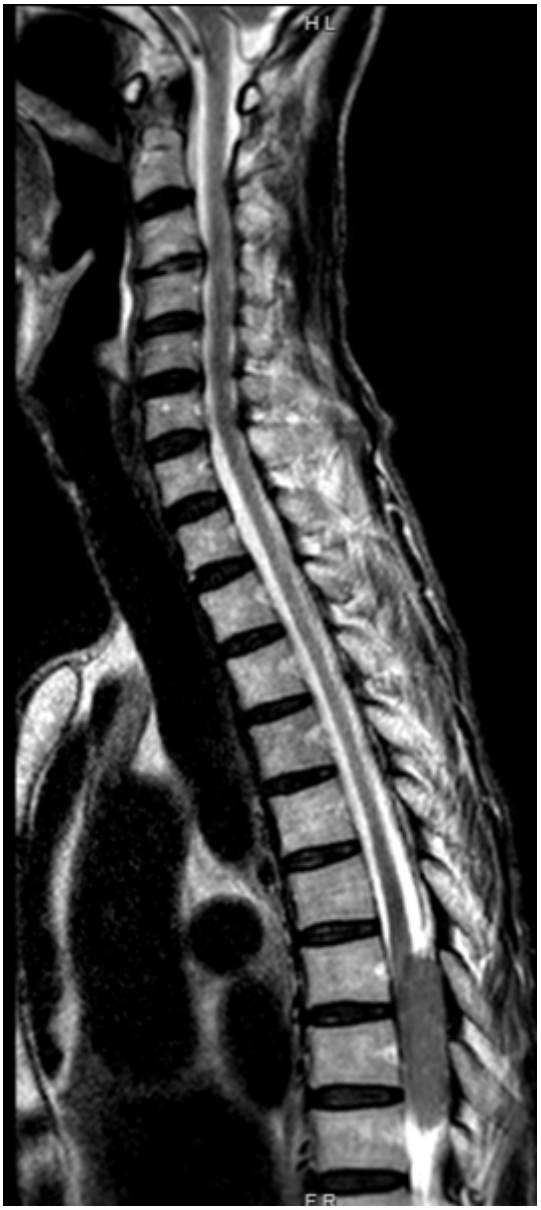
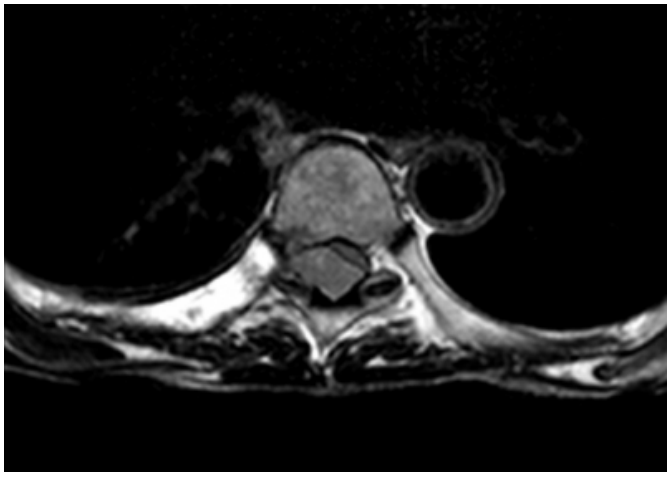


Figure 4

MRI Spine T2 showing a slightly hyperintense extramedullary lesion from T7 to T9 with compression on spinal cord.



Pathology of the tumour was plasma cell neoplasm as evidence by H&E section showing atypical plasma cells with marked nuclear pleomorphism, prominent nucleoli and mitotic activities. The atypical cells are diffusely positive for CD138 immunostaining which is a marker for plasma cells.

Figure 5

The atypical cells are diffusely positive for CD138 immunostaining which is a marker for plasma cells.

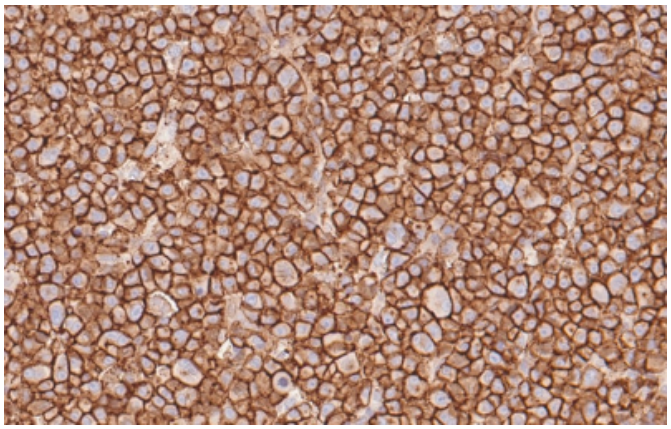
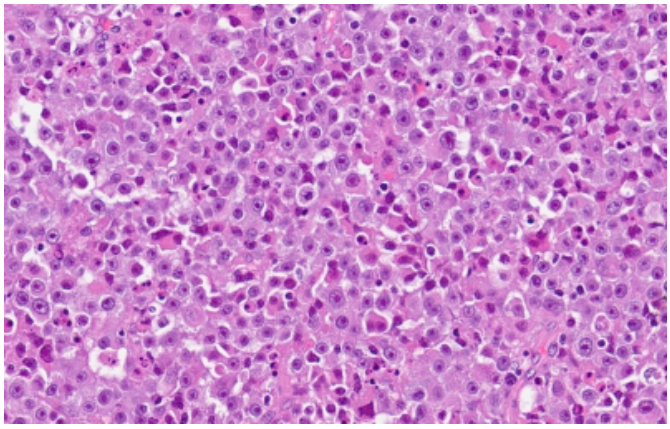


Figure 6

This is an H&E section showing atypical plasma cells with marked nuclear pleomorphism, prominent nucleoli and mitotic activities.



PET-CT showed no distant metastasis. He had 6% of clonal plasma cell in bone marrow examination and was considered as a case of solitary extramedullary plasmacytoma with minimal marrow involvement.

The patient is currently undergoing a course of rehabilitation. Unfortunately, his bilateral lower limbs power remained at 2/5 and he failed to wean off foley catheter post-operatively. He is now receiving palliative radiotherapy and thalidomide with multidisciplinary input from the medical and oncology colleagues.

DISCUSSION

The diagnostic criteria of solitary extramedullary plasmacytoma includes biopsy proven extramedullary tumour with clonal plasma cell, normal bone marrow with no evidence of clonal cell and no other lytic lesion in PET-CT scan and lack of end organ damage such as hypercalcemia, anemia, renal deficiency [3]. Solitary extramedullary plasmacytoma with minimal marrow involvement has similar diagnostic criteria exception its bone marrow contains less than 10% of clonal plasma cell.

In our patient, he had 6% of clonal plasma cell in bone marrow examination. He is considered as having a solitary extramedullary plasmacytoma with minimal marrow involvement. These patients are treated in the same manner with solitary extramedullary plasmacytoma, but they have a higher risk of progressing to multiple myeloma [4]. One study showed that half of the patient in this entity will develop multiple myeloma in two to three years [5].

Solitary spinal extradural plasmacytoma usually presents in MRI as spindle shape well defined lesion located in the

extradural dorsal spinal canal without bone erosion and paraspinal exophytic mass [6]. They have heterogeneously hypo or isointense signal on T1 weight imaging and hyper or isointense signal relative to muscle and gray matters on T2 weighted imaging. This can make the pre-operative diagnosis of Solitary spinal extradural plasmacytoma more difficult.

The morphologic features of extramedullary plasmacytoma include prominent nucleoli, easily seen mitotic figures and tumour giant cell. In some patient, 'Starry Sky' pattern was seen. A large proportional of extramedullary plasmacytoma has positive immunophenotype for CD138 (100%), CD38 (90.6%) and vs38c (95.5%)[7].

Treatment options of solitary spinal extradural plasmacytoma usually involve radiotherapy and surgical excision [6]. As it is an aggressive tumour, it is highly sensitive to radiotherapy. It is recommended for patient to receive fractionated radiotherapy with dose of 40-50 Gy involving at least one uninvolved vertebra [8].

Surgery is also usually performed aiming at decompression or diagnosis. Radiotherapy is still required after surgery as local recurrence rate was high in patient without adjuvant radiotherapy [9,10].

The role of adjuvant chemotherapy is controversial as results varies in different studies [11].

Some suggested that lenalidomide as an immunomodulatory agent plus dexamethasone can delay disease progression and improve patient survival according to the data for smouldering myeloma [10].

In our case, radiotherapy, thalidomide and dexamethasone were used with the expert opinion of hematologists and oncologists.

Conclusion

Solitary spinal extradural plasmacytoma is a rare entity of extradural tumour. It should be considered as a differential diagnosis if an extradural tumour is seen in the spinal canal without associated bone involvement. Treatment of the tumour required multidisciplinary approach from surgeons, oncologists and hematologists.

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