PRIMARY LEIOMYOSARCOMA OF SPINE – A RARE ENTITY

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ABSTRACT:

The primary leiomyosarcoma invading the spine is an extremely rare neoplasm .We report a case of 75 year old male with no significant medical history presenting with weakness in both lower limbs and incontinence of urine and faeces since 15 days. MRI confirmed the presence a lytic lesion causing collapse of D11-D12 vertebrae with involvement of paravertebral soft tissue and $11-12^{th}$ ribs. Decompressive surgery was performed for the same and tumour was sent for histopathological examination. On histology, tumour was found to be low grade leiomyosarcoma which showed positivity for desmin and smooth muscle actin on Immunohistochemistry.

Key words: leiomyosarcoma, primary, spine.

INTRODUCTION

Primary leiomyosarcoma (LMS) of the bone is a rare malignant neoplasm, accounting for only 0.64% of all bone tumours ¹.

It is generally assumed that primary LMS of bone arises from smooth muscle cells in the media of the intraosseous blood vessels.² In 1965, Evans and Sanerkin first reported extrafacial LMS of the bone.¹ The most frequently involved sites are the femur and tibia (around the knee) ^{3.} The vertebral column is an exceedingly rare location and only 13 cases have been reported until now⁴.

CASE REPORT

75 year old male with no significant medical history presented with past weakness in both lower limbs with incontinence of urine and faeces since 15 days. Neurological examination revealed grade I power in both lower limbs along with hypoesthesia below D12 level. MRI examination of thoracolumbar spine demonstrated a lytic lesion causing D11-12 collapse with involvement of paravertebral soft tissue and 11-12th rib. Complete radiological work up of the patient did not reveal any other tumour in the body.

D11-D12 decompressive laminectomy, D11-D12 partial corpectomy and D10-L1 transpedicular screw & rod fixation and stabilization was done and tumour was sent for histopathology. We received tumour in multiple tissue bits aggregating 3 cms, firm and brown in colour. Histology showed tumour composed of interlacing fascicles of spindle cells (figure 2) with eosinophilic cytoplasm with bluntended non-tapering nuclei (figure 3). Tumour was invading and destroying bony trabeculae (figure 1). 2-3 mitotic figures were noted per 10 high power fields. No areas of necrosis were seen. Tumour showed positivity for smooth muscle cell actin and desmin on IHC (figure 4&5). Based on these findings, the diagnosis of low grade leiomyosarcoma (LMS) was made. Since leiomyosarcoma metastatic are more common, complete radiological work up of the patient was done. Radiological work up did not reveal any other tumour in the body and thus. diagnosis of primary leiomyosarcoma of spine was made.

The postoperative period was uneventful and the patient's motor and sensory function improved, and he was discharged with recommendation of radiotherapy.



Figure 1: Tumour composed of spindle cells invading the bony Trabeculae (arrow) (H& E 100X)

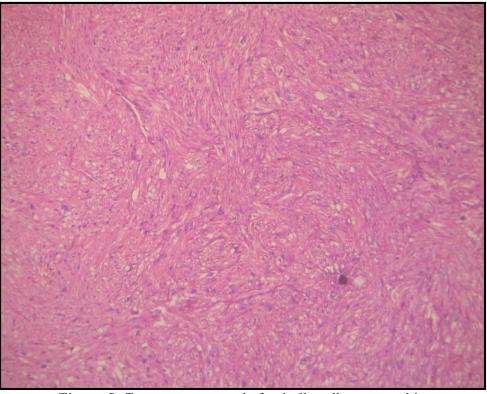


Figure 2: Tumour composed of spindle cells arranged in interlacing fascicles. (H&E 100X)

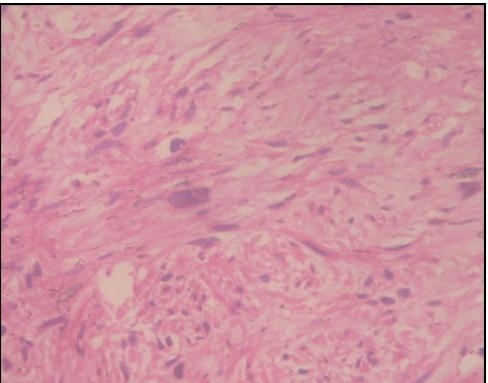


Figure 3: Spindle cells with eosinophilic cytoplasm and elongated nuclei with blunt end. Few hyperchromatic pleomorphic nuclei were noted. (H&E 400X)

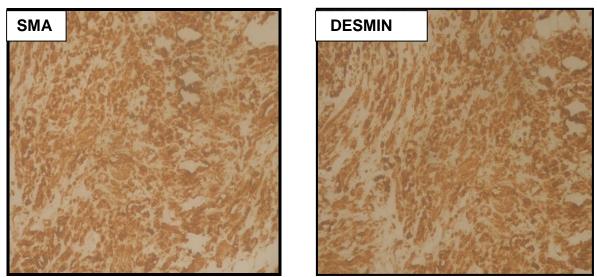


Figure 4 & 5: Tumour show positivity for smooth muscle actin (SMA) and desmin on immunohistochemistry (H&E 100X)

DISCUSSION

Leiomyosarcomas originate from smooth muscle cells and are relatively rare malignant tumours, accounting for 7 % of soft tissue sarcomas.⁵ They occur predominantly in the uterus and the gastrointestinal tract. Other common locations include the retroperitoneum and the subcutaneous tissue of the extremities.

The primary tumour subsequently metastasizes to distant sites, including the lung, liver, kidney, brain, and skin. Bone is rarely involved, usually as a late manifestation of the disease and the preferred location of bone metastasis is the spine¹. Primary LMS affecting the vertebral column is extremely rare and very few cases have been reported in the literature. Of the total seventeen primary cases of LMS of the spine, some were cases of LMS originating from the paravertebral soft tissue and not the vertebrae itself, presenting as extradural lesions compressing the spinal cord.⁶ To our knowledge, only 13 other cases of primary spinal LMS have been reported to date.⁴ The histological features of primary LMS of bone do not differ from those of LMS found elsewhere in the body.

Leiomyosarcoma of the spine may arise from intraosseous vascular smooth muscle cell ⁴, a multipotential mesenchymal stem cell or an intermediate cellular form (e.g., myofibroblast) capable of smooth muscle differentiation.² Paraspinal leiomyosarcoma arise from paraspinal musles, erodes spine and may involve nerve roots and major blood vessels. The common differential diagnosis of spindle cell neoplasms in the spine are Leiomyosarcoma, Malignant peripheral nerve sheath tumor, Cellular Schwannoma and solitary fibrous tumour. Therefore, the discrimination between primary and metastatic osseous involvement is only made by excluding an extra spinal primary site after a thorough workup. Total en bloc spondylectomy is the surgical treatment of choice, minimizing local recurrence.¹

CONCLUSION

Accurate histological typing of the spindle cell lesions of the spine is important from the treatment point of view. Primary leiomyosarcoma of spine should be kept in mind in differential diagnosis of spindle cell tumours of spine. Since metastatic tumours of the spine are more common than primary, a thorough work up of the patient should be done to exclude metastasis before labelling the tumour as primary.

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