

Case Report

Primary intraosseous malignant peripheral nerve sheath tumor of spine with a giant paraspinal and retrospinal subcutaneous extension

Ashis Patnaik, Sudhansu Sekhar Mishra, Satya Bhusan Senapati, Sunil Kumar Patra, Kalpalata Tripathy¹, Subrat Burma¹

Departments of Neurosurgery, ¹Pathology, SCB Medical College and Hospital, Cuttack, Odisha, India

E-mail: *Ashis Patnaik - dr_ash007@yahoo.co.in; Sudhansu Sekhar Mishra - drssmishra@gmail.com; Satya Bhusan Senapati - satya.bhusan.senapati@gmail.com; Sunil Kumar Patra - drsunil_patra@yahoo.co.in; Kalpalata Tripathy - kalpalata_tripathy@yahoo.co.in; Subrat Burma - drburma_subrat2010@rediffmail.com
*Corresponding author

Received: 23 August 12

Accepted: 23 October 12

Published: 26 December 12

This article may be cited as:

Patnaik A, Mishra SS, Senapati SB, Patra SK, Tripathy K, Burma S. Primary intraosseous malignant peripheral nerve sheath tumor of spine with a giant paraspinal and retrospinal subcutaneous extension. *Surg Neurol Int* 2012;3:157.

Available FREE in open access from: <http://www.surgicalneurologyint.com/text.asp?2012/3/1/157/105096>

Copyright: © 2012 Patnaik A. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Background: According to World Health Organization (WHO) classification of tumors, malignant peripheral nerve sheath tumors (MPNST) encompass the tumors, which were previously termed as malignant schwannoma, neurogenic sarcoma, and neurofibrosarcoma. These are rare tumors constituting only 5% of all malignant soft tissue tumors. As per their name, they arise from the malignant proliferation of cells forming sheath of a nerve root. They cause spinal cord compression, secondary changes in the surrounding bone with variable amount of tumor tissue going into the paraspinal space. However, purely intraosseous origin of the MPNST with no visible connection with a nerve root or dura is rare and few cases have been described in the literature.

Case Description: We present a primary intraosseous MPNST arising from the body of a thoracic spine with a minimal intraspinal component. However, there was a huge tumor part occupying the paraspinal and retrospinal region. The latter component was so large that it extended to lie just beneath the skin. The intraspinal component was confined to only one level. The giant extraspinal part was spanning multiple corresponding spinal level. We could not find such presentation in the literature.

Conclusion: Gross total removal (GTR) followed by adjuvant chemo-radiotherapy is the optimal treatment for MPNST of spine. In case of multiple laminectomy or gross spinal instability, spinal instrumentation makes the treatment protocol complete.

Key Words: Intraosseous, malignant peripheral nerve sheath tumor, paraspinal, spine

Access this article online

Website:
www.surgicalneurologyint.com

DOI:
10.4103/2152-7806.105096

Quick Response Code:



INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNST) usually arise from the peripheral nerves but are rare in comparison to their benign counterpart, neurofibroma or

schwannoma. They primarily affect adults in the third to sixth decades of life with an incidence of 0.001% in the general population.^[2] Primary intraosseous origin at spine level is extremely rare^[5,7] and its pathogenesis is unclear. Most probably, it arises by the proliferation of some of

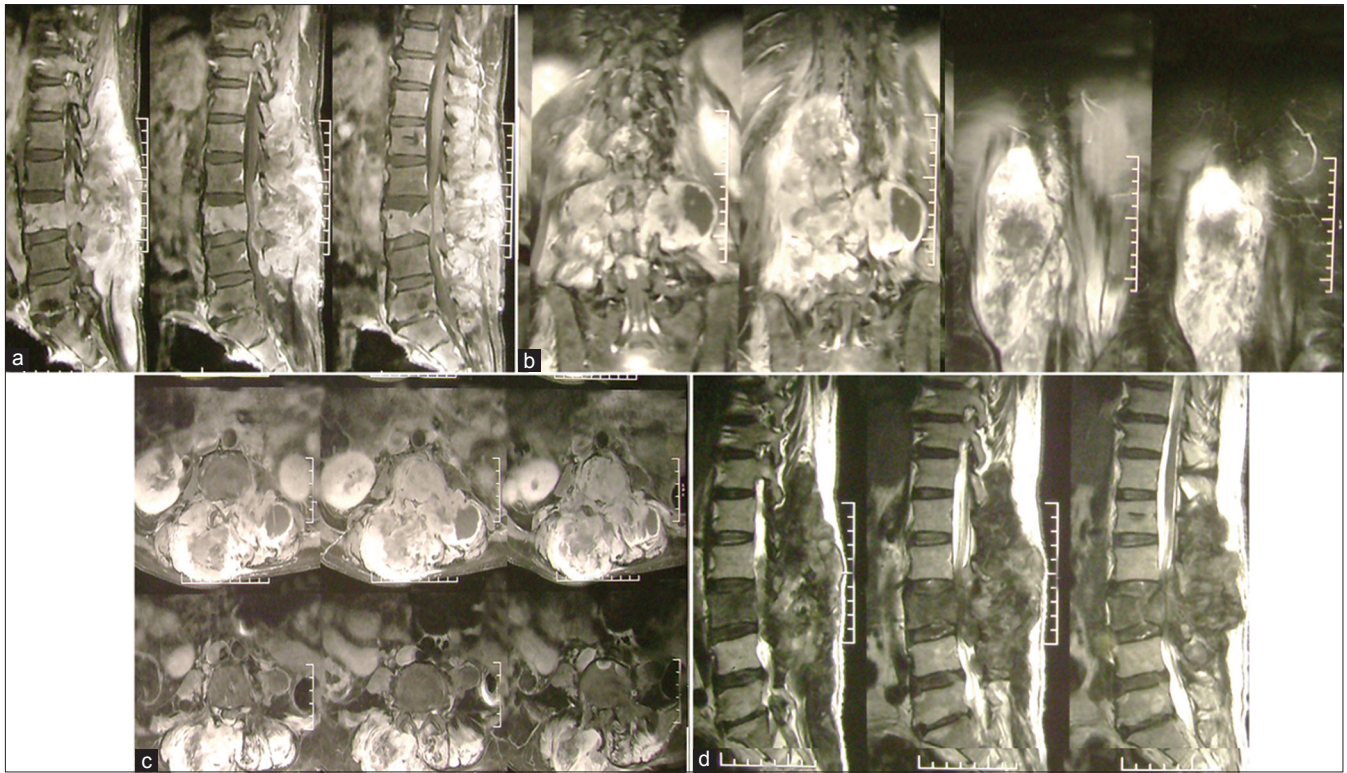


Figure 1: (a) Contrast sagittal image showing the L3 vertebral body collapse with a giant tumor component lying in retrospinal space. (b) Coronal contrast image showing extension of the tumor to both paraspinal spaces. (c) Axial contrast showing involvement of the L3 body with tumor coming out of the spinal canal through both side intervertebral foramina. Note there is no tumor at L4 intervertebral foramina. (d) T2 sagittal image

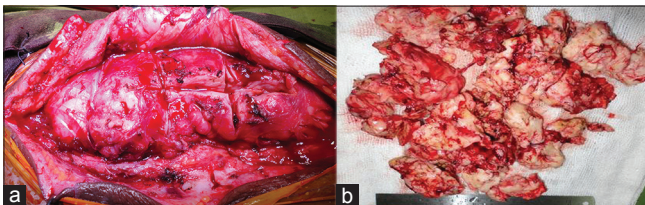


Figure 2: (a) Large extension of the tumor lying in subcutaneous plane. (b) Tumor specimen in piecemeal

the entrapped neural crest cells during vertebral body development.^[3] Review of literature fails to find the exact pathogenesis of such intraosseous nature of origin. The prognosis of these tumor is poor even with gross total resection due to their high malignancy and aggressive natural course with frequent recurrence and distant metastases. Literature shows that the tumor having paraspinal extensions have a more dismal outcome than their counterparts not having it.^[1,4] This may be due to the fact that in the former, chances of gross total resection becomes less due to extensive involvement of multiple intervertebral foramina and extraspinal vascular structures. In our case, single intervertebral foramen was involved with a giant component extending to paraspinal and retrospinal space. Primary intraosseous nature of the tumor was suggested by gross destruction of thoracic vertebral body, minimal intraspinal, intraforaminal

component, and nonattachment to a nerve root or dura. We did a palliative removal of the tumor, except the osseous part in view of limited life expectancy and the possible anesthetic and postoperative surgical complications in our severely morbid patient.

CASE REPORT

A 75-year-old male patient presented with rapidly progressive weakness of both lower limbs since past 3 months. At the time of presentation, he was bed ridden with grade 0 power in both lower limbs. Bladder involvement in form of urinary retention occurred 1 month back with patient on a indwelling Foley's catheter. There was a definite sensory level at L2 level for all modalities of sensation. Patient was known hypertensive and had ischemic heart disease with one episode of myocardial infarction and had undergone angioplasty 8 months back. Plain X-ray of lumbar region showed erosion of L3 vertebral body. Magnetic resonance imaging (MRI) pictures showed a large mass lying in paraspinal and retrospinal region [Figure 1]. The retrospinal extension was so massive that it almost extended to lie just beneath the skin. There was a small intraspinal component compressing the cord. L3 vertebral body was collapsed with tumor seen communicating into it. The intraspinal component extended outside only through

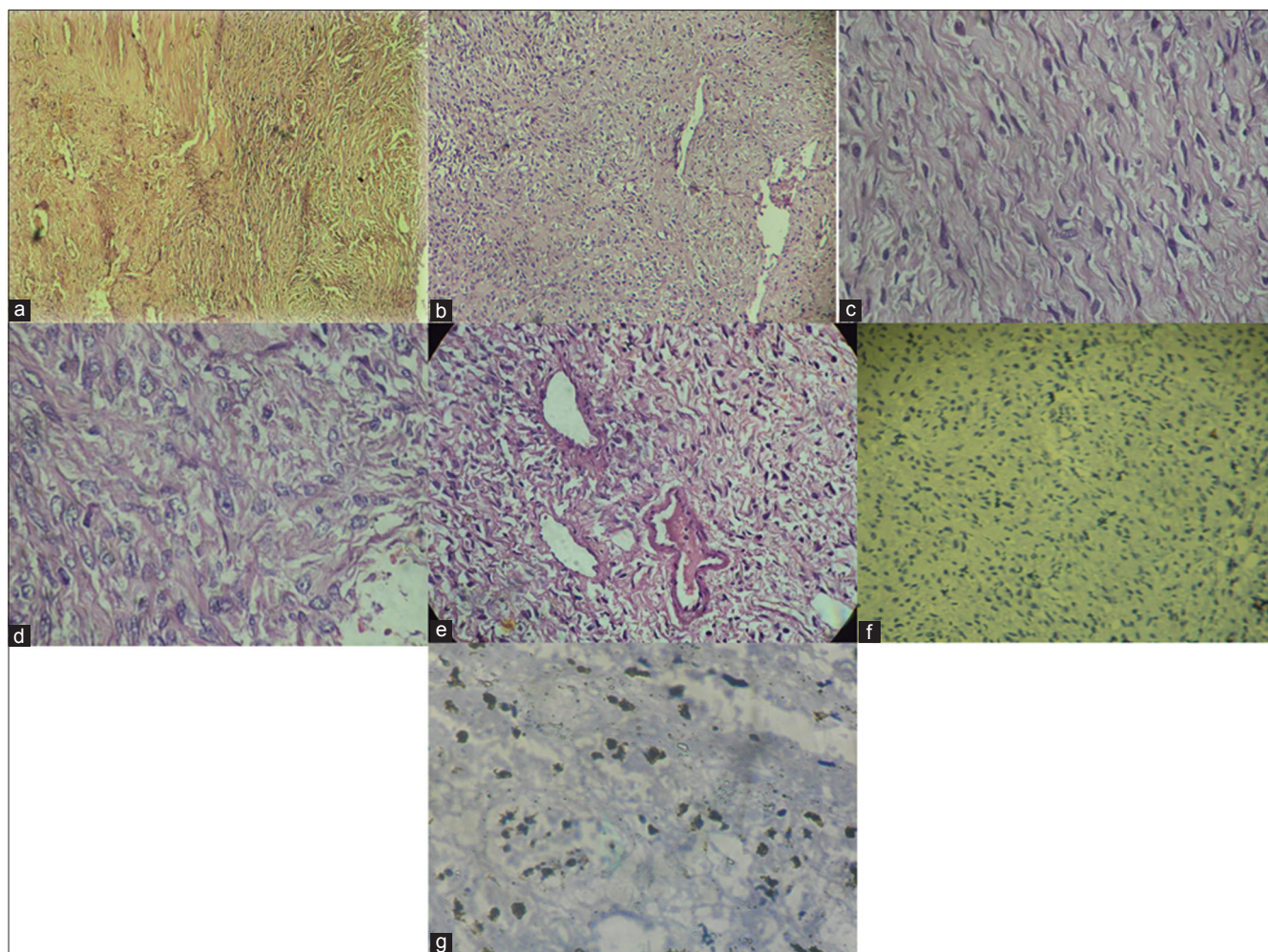


Figure 3: (a) Densely cellular fascicles alternating with hypocellular myxoid zones (x200). (b) Spindle shaped cells in a vague storiform pattern (x200). (c) Cells have irregular contours with eosinophilic cytoplasm with wavy, buckled, comma-shaped nuclei (x400). (d) Cells showing pleomorphism, prominent nucleoli (x400). (e) Perivascular accentuation of tumor cells (x200). (f) Negative GFAP immunostaining. (g) Strong Ki-67 immunopositivity (x400)

one intervertebral foramen. However, the extraspinal component was seen extending several corresponding spinal levels from L1 to L4. The metastatic workup for the patient was negative. Another location of the tumor in the body could not be found out even after thorough work-up.

A linear incision given on the back from L1 to L4 spine. The tumor was found to lie just beneath the skin [Figure 2a]. The extraspinal part of the tumor was removed. The tumor was reddish brown, fleshy, vascular [Figure 2b] with infiltration into the surrounding tissue. L3 laminectomy was performed and the intraspinal part excised. The tumor did not have any obvious connection with any nerve root or dura. There was no plane of demarcation between the tumor and the vertebral body. Vertebral body excision, grafting, and fusion procedures were not done because of gross of co-morbidities with anesthetic risk.

Histopathology showed densely cellular fascicles alternate with hypocellular myxoid zones [Figure 3a]. The cellular areas showed large number of spindle

cells arranged in a storiform pattern [Figure 3b]. Each of the spindle cells have irregular contours with abundant eosinophilic cytoplasm and gross cellular atypia, mitotic figures, pleomorphism. Nuclei were wavy, buckled, comma shaped [Figure 3c] with prominent nucleoli [Figure 3d] suggestive of malignant variety of peripheral nerve sheath tumor. Histopathological evaluation was done by two independent pathologists and their impression were same. There were occasional areas of epithelioid differentiation and perivascular accentuation of tumor cells [Figure 3e]. Negative glial fibrillary acidic protein (GFAP) immunohistochemistry ruled out astroglial origin [Figure 3f]. Ki-67 proliferative index was 5.5%, suggestive of malignant nature of the tumor [Figure 3g].

DISCUSSION

MPNSTs are a rare variety of soft tissue sarcomas arising from proliferation of schwann cells. These tumors have

been described to have multiple cell line origins. Schwann cells are neural crest derivatives. Primary intraosseous nature of schwann cells could be explained on the developmental entrapment of some of the neural crest cells into the vertebral body, which differentiate along the schwann cell lineage. Later malignant proliferation of this isolated group of cells could have given rise to the MPNST of the present case. Primary intraosseous nature of the tumor is supported by a small intraspinal component, nonattachment of the tumor to any nerve root intraoperatively. MPNSTs cause secondary changes in the vertebral body and spine. But gross collapse of the body as in the present case is more suggestive of intraosseous lesion than secondary changes. The tumor was coming out of the spinal canal only through a single intervertebral foramen and was causing gross bony destruction. It rules out the possibility of a primary mediastinal MPNST secondarily entering the spinal canal. Few such cases of primary intraosseous MPNST have been described in the literature.^[2]

Clinico-radiologically the mass appeared to be a highly aggressive lesion due to its short history, huge size, infiltrative nature. Possibility of neurofibroma was low because of very short history and bony destructions. Differential diagnoses include sarcomatous growths, metastatic deposits, neuroectodermal tumors, and ganglioneuroblastoma.

Histologically MPNST belongs to spindle cell group of tumors. It can be differentiated from other sarcomas such as synovial sarcoma by the presence of cells showing neuronal differentiation and presence of typical cyto-nuclear features such as abundant eosinophilic cytoplasm and wavy, buckled, comma-shaped nuclei. Immunohistochemistry only helps in ruling out tumors of astrocytic, meningothelial origin. S-100 is positive similar to any other tumor of neuronal origin. Other markers such as Leu-7 and major basic protein are less sensitive in detecting MPNSTs. These tumors show high proliferative indices suggestive rapid growth and a poorer outcome due to early recurrences and distant metastases.

We achieved the complete removal of soft tissue part of the tumor. Removal of the osseous part of the tumor by corpectomy of the involved vertebrae through thoracotomy followed by grafting and instrumentation would have made the best treatment for the patient. This would have made the operative procedure lengthy with associated postoperative pulmonary, graft and instrumentation-related complications. Such complications have been reported in the literature after gross total resection of MPNST in a young female.^[6]

MPNST of spine is itself rare. Primary intraosseous nature along with such a huge paraspinous and retrospinous extension makes our case exclusive. Although we could achieve a palliative excision of tumor, complete excision including the osseous part followed by adjuvant radio-chemotherapy is the best treatment. In spite of this, as the outcome is still poor, hence further studies on genetic therapy of the tumor based on its molecular pathogenesis is required.

REFERENCES

1. Ducatman BS, Scheithauer BW, Piepgras DG, Reiman HM, Ilstrup DM. Malignant peripheral nerve sheath tumors. A clinicopathologic study of 120 cases. *Cancer* 1986;57:2006-21.
2. Gnanalingam K, Bhattacharjee S, O'Neill K. Intraosseous malignant peripheral nerve sheath tumor (MPNST) of the thoracic spine: A rare cause of spinal cord compression. *Spine (Phila Pa 1976)* 2004;29:402-5.
3. Hajdu SI. Peripheral nerve sheath tumors. Histogenesis, classification, and prognosis. *Cancer* 1993;72:3549-52.
4. Jandali S, Diluna ML, Storm PB, Low DW. Use of the vascularized free fibula graft with an arteriovenous loop for fusion of cervical and thoracic spinal defects in previous irradiated pediatric patients. *Plast Reconstr Surg* 2011;127:1932-8.
5. Khan RJ, Asgher J, Sohail MT, Chughtai AS. Primary intraosseous malignant peripheral nerve sheath tumor: A case report and review of the literature. *Pathology* 1998;30:237-41.
6. McLaughlin EJ, Heuer GG, Whitmore RG, Birknes JK, Belasco J, Sterman D, et al. Treatment of a malignant peripheral nerve sheath tumor and its complications through a multidisciplinary approach. *J Neurosurg Pediatr* 2011;7:543-8.
7. Miyakoshi N, Nishikawa Y, Shimada Y, Oada K, Enomoto K, Itoi E. Intraosseous malignant peripheral nerve sheath tumor with focal epithelioid differentiation of the thoracic spine. *Neurol India* 2007;55:64-6.