

Case Report

A rare case of primary extranodal, extradural histiocytic sarcoma of the thoracic spine and review of literature

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Abstract

Background: Histiocytic sarcoma (HS) is a rare aggressive malignant neoplasm of hematopoietic cell origin showing features of fever, weight loss, adenopathy, hepatosplenomegaly, and pancytopenia. Primary histiocytic sarcoma of spine without involvement of lymph nodes, bone marrow or systemic features is very rare. Due to highly malignant, aggressive behavior and frequent local and distant metastases, prognosis of histiocytic sarcoma is poor.

Case description: We report a case of primary extranodal variety of histiocytic sarcoma without involvement of bone marrow at extradural location of thoracic spine. Immunohistochemistry panel ruled out other more common lesions in this location. We could not find such presentation of extradural histiocytic sarcoma at the thoracic region in the literature.

Conclusion: Primary involvement of extradural space by histiocytic sarcoma could arise from the proliferation of multipotent hematopoietic stem cells which are left in this space embryologically.

Key Words: Extranodal, histiocytic sarcoma, primary, spine

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INTRODUCTION

Histiocytes are the antigen-processing phagocytes derived from hematopoietic or mesenchymal stem cells. Along with dendritic cells, they are involved in antigen presentation, processing, and final destruction of antigens through phagocytosis. Histiocytic sarcoma (HS) is a rare hematopoietic neoplasm with aggressive behavior and poor outcome. It can present as localized disease confined to the skin, lymph nodes, and intestinal tract, or as disseminated disease.^[2] Our case was a young male patient presenting with short-duration paraplegia with bladder involvement. The radiological picture

was of infiltrative growth at the extradural location of thoracic spine suggestive of more common hemangioma or metastatic disease. The intraoperative picture was of malignant lesion due to the vascular, friable nature of the lesion. Histopathology was suggestive of a malignant neoplasm of histiocytic origin. Immunohistochemistry confirmed the lesion to be histiocytic sarcoma. There was only one such presentation of isolated extradural primary histiocytic sarcoma of spine in the literature.^[1]

CASE REPORT

A 22-year-old male patient presented with a 3-month

history of both lower limb weakness progressing rapidly to paraplegia. There was associated burning pain in the same area. The patient was catheterized due to urinary retention 2 months back. There was no associated fever, lymphadenopathy or significant weight loss. On examination both the lower limbs were hypertonic with gross spasticity. Motor power was grade 0 (MRCS scale) around all the joints with the patient being bed ridden with trophic ulcer at the sacral region. Deep tendon reflexes were exaggerated with bilateral patellar and ankle clonus. There was a specific sensory level at T10 below which all modalities of sensation were impaired.

Plain x-ray was nonspecific. On MRI, there was a grossly infiltrating lesion at the level of T8, T9 and partially extending to T10 level [Figure 1a and c]. Signal changes were also noted in the T8, T9 vertebral bodies. The lesion was extradural compressing the cord all around like a cylinder which was well seen in the axial cut [Figure 1b]. The lesion had an irregular margin with intense homogenous enhancement on the gadolinium contrast image. Routine blood investigations, bone marrow study, bone scan, chest x-ray, ultrasound abdomen, and pelvis were normal.

T8, T10 laminectomy was done. Just beneath the laminae a vascular, pinkish white, friable, soft mass found [Figure 2] completely carpeting the cord from all sides. It was nonadherent to the cord and was easily separated from it. The posterior and lateral part of the mass was removed. Component anterior to cord could be removed by partial medial facetectomy at T8 and T9 level. Part of the tumor was seen entering intervertebral foramen at T8 level which was removed piecemeal.

Histopathology showed sheets of large, pleomorphic, mononuclear cells showing marked cytologic atypia admixed with large no of lymphocytes [Figure 3a]. Cells were large epithelial like with abundant pale eosinophilic cytoplasm with oval to irregular nuclei,

vesicular chromatin, prominent nucleoli with numerous mitotic figures [Figure 3b]. There were binucleate and multinucleated giant cells [Figure 3c]. Few areas showing phagocytosis of red cells, leukocytes, and tumor cells by the histiocytes called emperipolesis [Figure 3d] were seen. Immunohistochemistry typically showed positivity toward S-100 [Figure 4], CD68 [Figure 5], and particularly lysozyme [Figure 6] and was negative for CD1a, CD21, langerin suggesting the final diagnosis as histiocytic sarcoma. Staining for myeloperoxidase, CD34, and CD117 were negative excluding myeloid sarcoma. Trepine biopsy of bone marrow from iliac crest showed no tumor cells confirming the isolated involvement of spine.

After 30 days postoperatively, the patient's power in both lower limbs improved to grade 2. The patient refused to take both radio- and chemotherapy and is alive 5 months after surgery without any recurrence.

DISCUSSION

Although histiocytic sarcoma is a neoplasm of hemolymphoid cell lineage, frequently it arises in nonlymphoid organs. However its primary occurrence in spine causing its compression is very rare and few cases were reported in the literature very recently.^[1,3,4] It is a highly malignant neoplasm showing frequent areas of atypia, mitotic figures, nuclear pleomorphism. Corresponding to its aggressive behavior it usually has a short duration of symptoms as shown in the present case. Usually the spine involvement results in rapidly progressing paraparesis leading to complete paraplegia. Bowel, bladder involvements are frequent. Histiocytic sarcoma can present as a disseminated disease with involvement of multiple organ systems such as fever, weight loss, adenopathy, hepatosplenomegaly, and pancytopenia. Sohn *et al.* reported a case of histiocytic sarcoma

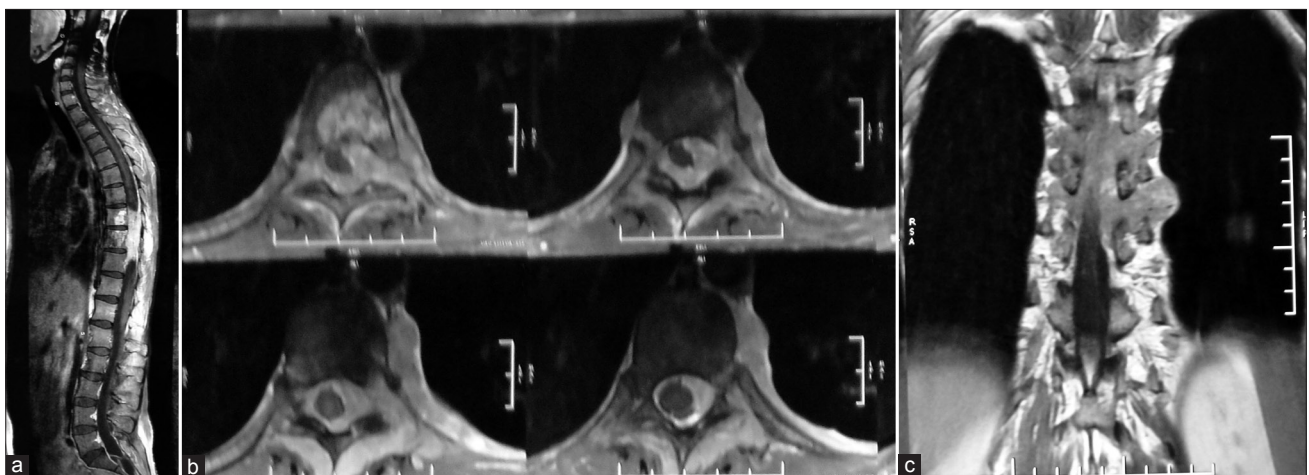


Figure 1: (a) T1-weighted contrast sagittal image showing the tumor with irregular margin and intense enhancement on contrast. Bodies of T8 and T9 vertebrae show infiltration. (b) T1-weighted contrast axial image showing the enhancing tumor surrounding and compressing the tumor from all side like a cylinder. (c) Coronal image showing the infiltrative nature of the tumor

presenting with primary bone marrow involvement with systemic symptoms, including fever, weight loss, and

generalized weakness.^[4] However in the present case, spine involvement was isolated without any systemic features including the negative bone marrow study.

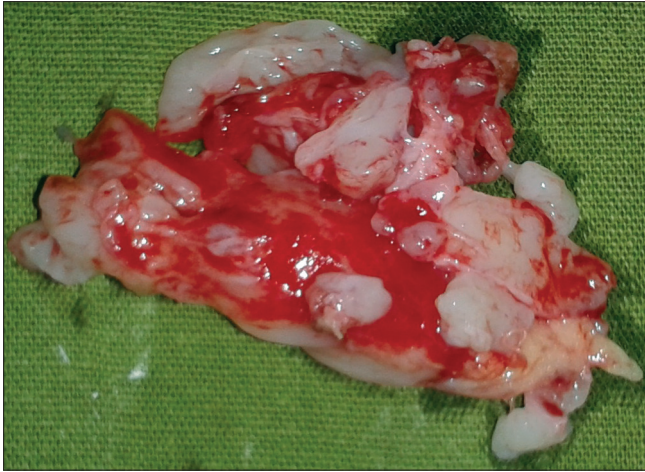


Figure 2: Tumor specimen showing the pinkish white color, soft, friable nature of the tumor

Radiologically histiocytic sarcoma is quite difficult to diagnose as it frequently presents as infiltrative lesion with laminae, spine, and body erosion suggestive of more common metastatic lesion. It may be located in the extradural location as in the present case. Kaushal *et al.* reported a case located at the lumbar extradural region.^[1] However, primary intramedullary histiocytic sarcoma has been reported in the literature.^[4] In the present case the extradural was eroding the spine, laminae and infiltrating the body of T8 and T9 vertebrae, but nonadherent to the dura. Differential diagnosis would include metastatic lesion, tuberculosis of spine, hemangioma with vertebral body involvement with intraspinal extension.

Diagnosis of rare histiocytic sarcoma is purely histological with large numbers of pleomorphic, large epithelial like

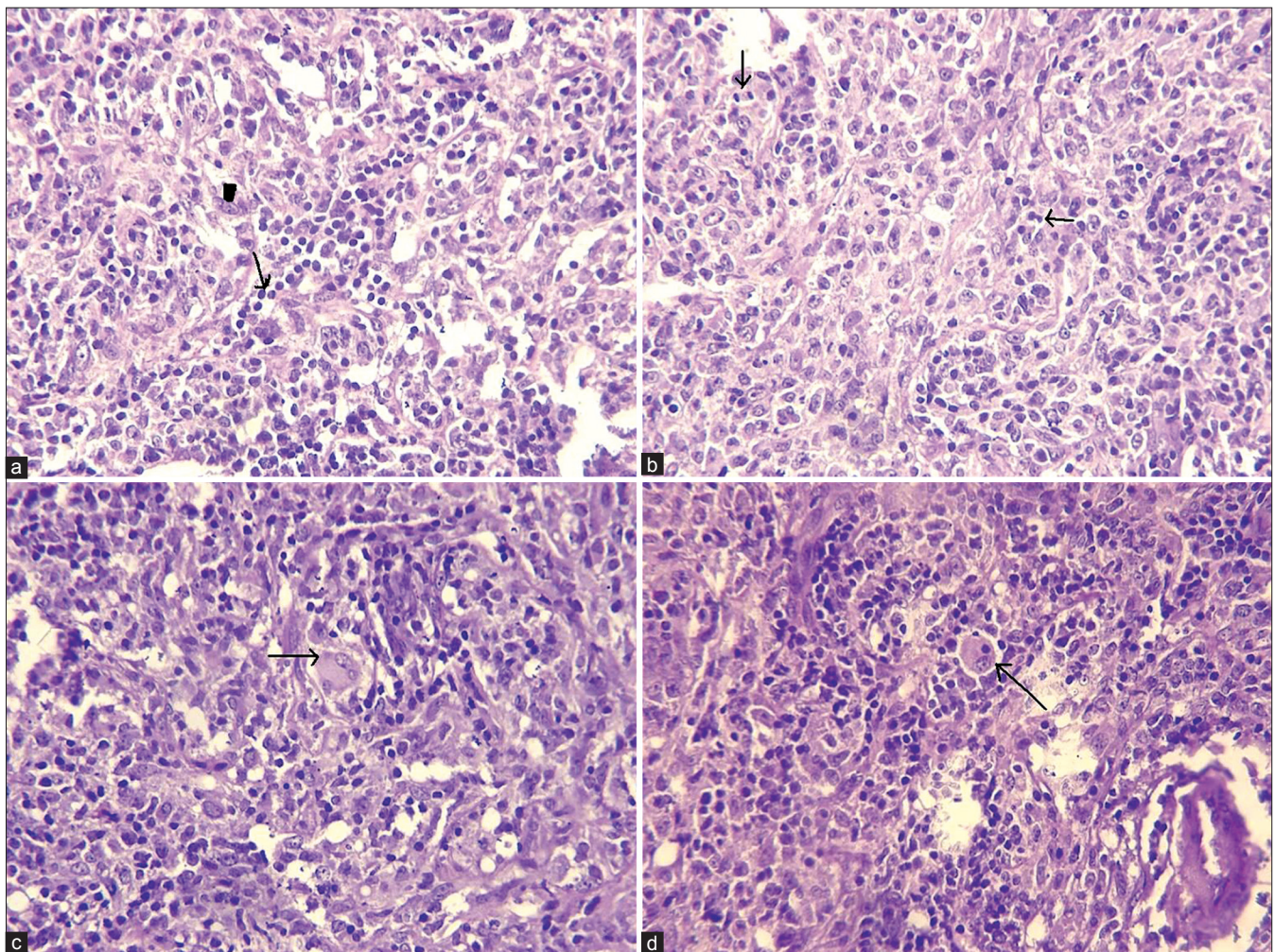


Figure 3: (a) Routine staining show pleomorphic, large cells with abundant cytoplasm (arrowhead) with large numbers of lymphocytic infiltrate (long arrow). (b) Numerous mitotic figures (arrow). (c) Tumor giant cells (arrow). (d) Emperipolesis: Phagocytosis of red cells, leukocytes and tumor cells by the histiocyte

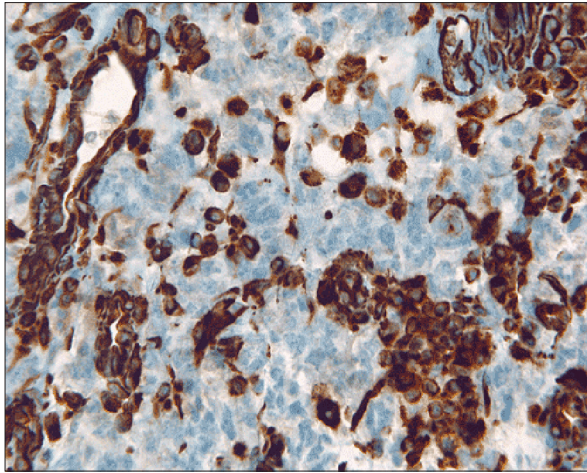


Figure 4: Immunohistochemistry with S-100 positivity

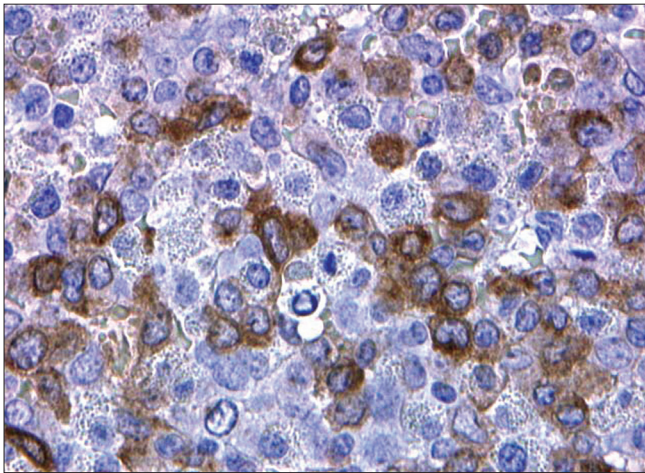


Figure 6: Lysozyme immunopositivity

cells with abundant cytoplasm and showing frequent areas of mitoses. These cells are the neoplastic histiocytes. Differentiation from histiocytic sarcoma from Rosai Dorfmann disease is difficult on plain histology with later showing more frequent cells containing phagocytosed red cells, leukocytes, and tumor cells by the histiocytes called emperipolesis. Immunohistochemically, both show positivity for CD68, but lysozyme is specific for histiocytic sarcoma. CD1a is positive in Langerhans histiocytosis, whereas it is negative in both histiocytic sarcoma and

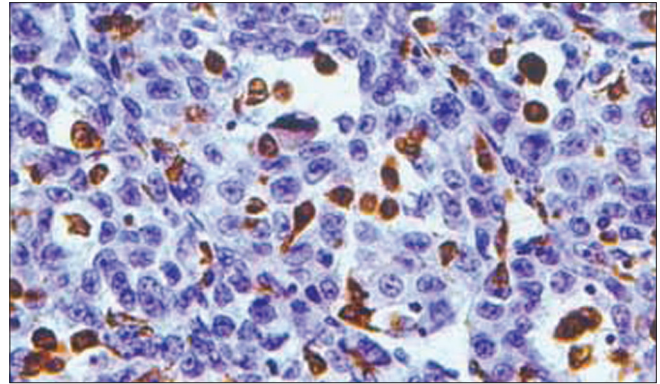


Figure 5: CD68 immunopositivity

Rosai Dorfman disease. Other most important differential diagnoses which need to be addressed are obviously a lymphoma and a myeloid sarcoma.

Surgical excision is the gold standard treatment for this rare lesion. However, as very few cases have been reported till now, the natural history as well as role of chemo- and radiotherapy is not clearly known. In the present case near total excision was done and the patient refused to undergo further adjuvant therapy.

Histiocytic sarcoma is a rare lesion and in recent years few cases was reported as a cause of spinal cord compression. Its diagnosis preoperatively is not possible and good histopathological study with immunohistochemistry clinches the diagnosis. In the future with reporting of more cases of histiocytic sarcoma, natural history as well as optimum treatment protocol for these rarer lesions could be carved out.

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