Malignant giant cell tumor of soft parts in lumbosacral region

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Abstract

Background: Giant cell tumor of soft parts is a rare neoplasm that mainly affects adulst and the elderly and is usually located in the extremities. Here we report a child with giant cell tumor of soft tissue, which is a very rare condition in childhood.

Clinical presentation: A 5 year old girl presented with a 5 month history of left lower extremity pain. She had developed paraplegia before admission. On examination, mild left lumbosacral swelling and tenderness was found. Abdominal and pelvic CT-Scan revealed an expansile lytic lesion of the left side of sacrum with significant soft tissue component extending toward the left iliac bone. Lumbar MRI revealed a space occupying lesion originating from posterior L5 elements, projecting toward the L1.

Intervention: The patient underwent surgery. A firm epidural hemorrhagic tumor of L5, S1, and S2 with no spinal cord involvement was found. Partial tumor resection (measuring 3*1*0.5 cm in maximal diameter) and laminectomy was done.

Conclusion: Primary giant cell tumors of soft tissue are distinctive, rare neoplasms that exhibit a wide clinicopathologic spectrum similar to osseous GCTs and need to be differentiated from other giant cell rich soft tissue tumors. Recognition of this tumor is important due to its behavior as a low grade malignancy, but this cannot be predicted and metastasis does occur rarely.

Keywords: Giant cell tumor of soft tissue (GCT-ST), large cell sarcoma of tendon sheath.

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Introduction

Giant cell tumor of soft tissue (GCT-ST) is a relatively rare entity, and is clinically and histomorphologically indistinguishable from its counterpart in bone (3). It was first described in 1972 by Salm and Sissons, followed shortly by Guccion and Enzinger (4). It affects adults of both sexes (3). It seldom occurs in the first two decades of life (1). Most cases are located deeply, but a superficial variety in subcutaneous tissue and fascia has also been described. (2)

The tumor is composed of an admixture of osteoclast like multinucleated giant cell and stromal cells. The stromal cells are probably the only neoplastic component, some are elongated and fibroblast like, whereas others are plump, resembling histiocytes. In earlier schemes this tumor had been included as one of the histologic types of MFH, but this is no longer favored (2). The behavior is dependent upon the location, size and microscopic appearance. Low grade (benign, of low malignant potential) and high grade (malignant) forms have been separated from each other on the basis of the atypia, pleomorphism and mitotic activity of the mononuclear neoplastic component. (2)

It is treated adequately by complete excision; a benign clinical course is expected. The episodes of distant metastasis and tumor associated death seem to be exceedingly rare.

Case report:

The patient was the 5 year daughter of nonconsanguineous, healthy parents, with the family history of congenital spinal tumor in her mother's cousin. She had no history of perinatal complications, but febrile convulsion in infancy and mumps 7 month prior to admission. She presented with spontaneous left lower extremity (leg and sole) pain. Medical analgesic therapy and physiotherapy was done, but she had developed sitting disabilities, claudication, lumbar pain, fever and anorexia.

On examination, mild left side lumbosacral swelling and tenderness was found. No other remarkable finding was detected in physical examination.

Multidetector abdominal and pelvic CT-Scan with oral and IV contrast revealed an expansile lytic

lesion of left side of sacrum with significant soft tissue component extending toward the left iliac bone. Visible bony structures, psoas and paravertebral muscles were in normal limits. MRI revealed a lesion originating from L_5 toward left sacrum. Lumbosacral radiography, lung and brain CT-Scan were unremarkable.

The patient underwent surgery. An epidural hemorrhagic tumor of L_5, S_1, S_2 with no spinal cord involvement was found. Partial tumor resection and laminectomy was done. Microscopic examination revealed a malignant neoplasm composed of numerous multinucleated giant cells embedded in a stroma with atypical pleomorphic spindle cells and some histiocytes showing moderate mitotic indices including a few atypical mitotic figures. (Figures 1 and 2)

The neoplasm invaded adjacent bone, entrapping bony sequestrate. Bone marrow aspiration revealed hypercellularity with increased myeloid series and plasma cells. Bone scan showed bony lesion in lateral aspect of the upper sacral region (left sacroiliac joint). The patient was discharged on the 9th day of admission.

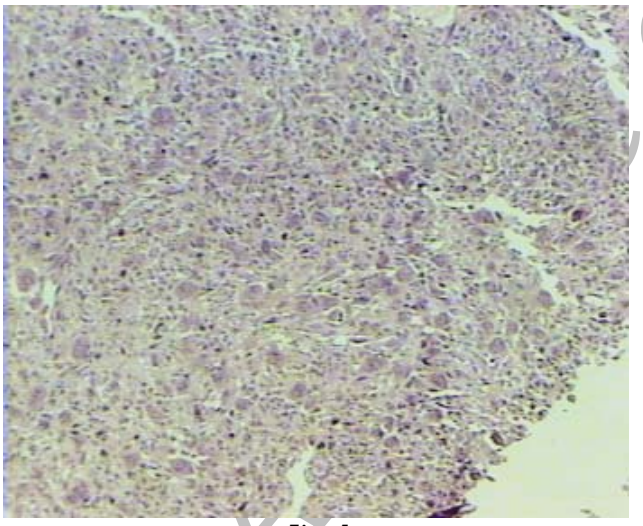

Fig. 1

Discussion:

Primary giant cell tumors of soft tissue are distinctive neoplasms that, like osseous giant cell tumors, exhibit a wide clinicopathologic spectrum. These neoplasms should be distinguished from other giant cell-rich soft tissue tumors with which they may be confused. The differential diagnosis includes malignant fibrous histiocytoma, extra osseous osteosarcoma, epithelial sarcoma and fibrosarcoma (1).

Today, it is considered as the soft tissue analogue of giant cell tumor of bone according to light microscopic, ultrastructural and histochemical characteristics (7). The tumor capacity to recur or to evolve into a malignant lesion is recognized, but with exceedingly low recurrence rate, provided the lesion is removed adequately (5, 6). In malignant GCT-ST, there is histological evidence of sarcomatous changes as indicated by cellular and nuclear pleomorphism, areas of hemorrhage, necrosis with high mitotic activity of mononuclear cell (MNC) histiocytes and fibroblasts.

The MNC in benign lesions are devoid of atypia, pleomorphism and atypical mitoses but no histopathologic feature is pathognomonic of aggressive behavior of the tumor.

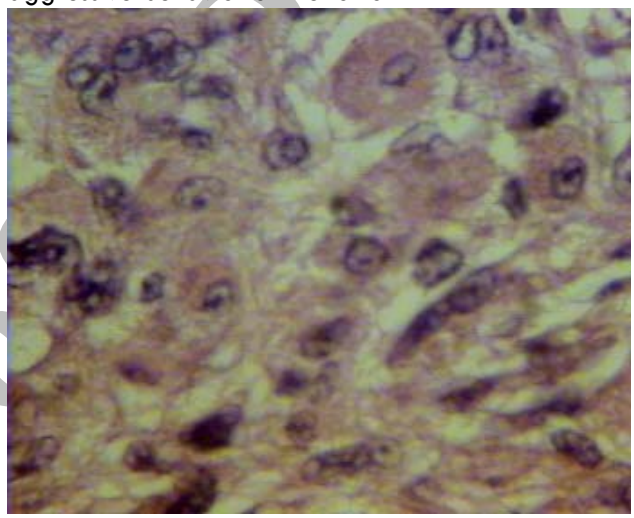

Fig. 2

An average number of mitotic figures in benign GCT-ST varies from 2-3/10 Hpf in one series to 9.5/10 Hpf in another series where 6 out of 22 tumors revealed vascular invasion but these tumors did not metastasize (5). Metaplastic bone may be present, usually in from of peripheral shell (2). We found histiocytes, atypical pleomorphic spindle cells, and moderate numbers of mitotic figures with few atypical forms and also invasion to adjacent bony tissue in our case. MGCT of soft parts (large cell sarcoma of tendon sheath) seldom occurs in the first two decades of life (1), and most are deeply located in the extremities, but a superficial variety in the subcutaneous tissue and fascia has also been described. (2)

Almost all reported cases were in adults, there are reports of tumors occurring primarily in the dermis (9). A case has been reported in the paravertebral region opposite D3-D5 (10). Verhagen Wim et al, reported a 24 year old woman with monoradicular pain in the right leg in the region of S_2. Neuroradiologic examination showed a mass within the sacral spinal canal comprising the right S_2 root,

with no sign of bone involvement. Sacral laminectomy was performed and a soft tissue giant cell tumor was identified in histologic examination. The patient recovered completely. (11)

Flope et al, reported 31 cases of soft tissue GCT with mild to moderate nuclear atypia and proposed the entity as GCT of low malignant potential (8). Col Kailash et al, reported 7 cases of GCT-ST, all were superficial, circumscribed and involved extremities except one. (3)

Prognosis of GCT-ST varies and biological aggressive course for its local recurrence cannot be predicted. Proper surgical excision and a long period of follow up are essential in these cases (3). Our patient underwent surgery. As the tumor was an epidural hemorrhagic mass of L_5, S_1, S_2 with adjacent bone involvement, partial tumor resection and laminectomy was preferred and chemotherapy was considered.

In conclusion, this is the first description of a child with an epidural GCT-ST of lumbosacral region with secondary involvement of adjacent bony structures.

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