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Soft Tisue Giant Cell Tumour of Low Malignant Potential – A Rare Mediastinal S.O.L

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Abstract: Giant cell tumour of soft tissue (GCT-ST) particularly in posterior mediastnum is a very rare tumour. A seventeen year female who presented with superior vanacaval syndrome had a posterior mediastninal S.O.L. Thoracotomy was done and the mass was excised. Histopathological and immunohistochemical examination proved it to be a Soft Tissue Giant Cell Tumour of low malignant potential. Patient received E.B.R.T and is doing well till date. This study highlights rarity of the lesion and draws attention of its uneventful clinical course after complete excision.
Keywords: GCT-ST –Giant Cell Tumour Of Soft Tissue, Mediastinum.

INTRODUCTION

"Soft tissue giant cell tumors of low malignant potential" was proposed for a group of lesions that represent the benign end of the spectrum of malignant giant cell tumor of soft parts and that seem to be the soft tissue analogue of giant cell tumor of bone.

The nodules are composed of bland mononuclear cells, short spindle cells and osteoclast-like giant cells. The majority of these tumors have been reported to occur in the lower extremities. Giant cell tumor of soft tissue (GCT-ST) especially in the posterior mediastinum is a very rare tumor.

CASE REPORT

A seventeen year female was having persistent headache and swelling of face for three months before admission. The symptoms were gradually progressive. She also had episode of fever and vomiting. Physical examination revealed anaemia, facial puffiness, engorged veins in neck and upper extremity. There was diminished breath sound in right upper part of chest mostly in posterior aspect. C.T. showed a mediastinal S.O.L. in right upper hemithorax (Fig.1a). FNAC was inconclusive. After proper preoperative preparation and routine investigations, a plan of thoracotomy and exicision of tumour was done.

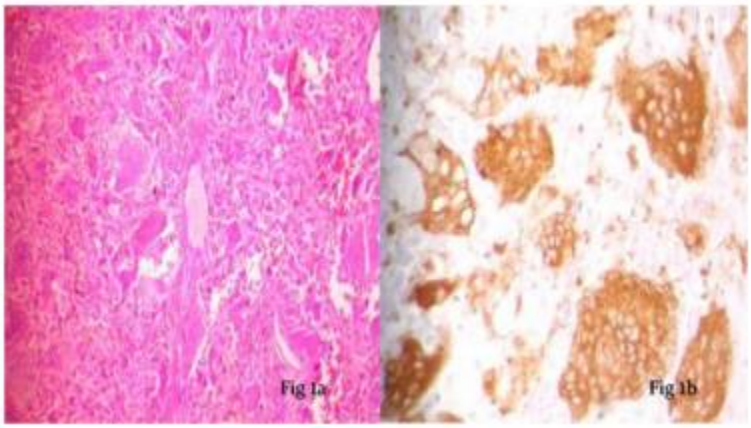

Fig:1a Photomicrograph showing osteoclast like giant cells distributed against a background of round to oval stromal cells (10x10 H&E).Fig 1b: showing CD68 positive giant cells

On right thoracotomy ,a huge vascular mass was seen in right upper mediastinum The mass was adherent to lungs, chest wall and azygos vein. The adhesions were dissected and the was excised totally.

Recovery was uneventful and the symptomatology improved.

Postoperatively she received External Beam Radiotherapy (E.B.R.) and till now she is doing well and on regular follow up. The original tumour was a spherical mass about 9cm. in diameter. The mass was highly vascular and friable, so it was excised in pieces. The fragmented tissue pieces were greyish white in colour, partly soft and partly firm in consistency. Sectios were prepared from different portons of excised mass and routine H&E stains were done followed by immunohietochemistry. Microscopically, the tumour was composed of sheets of mononuclear that that blended with spindled cells and benign osteoclastic giant cells (Fig.-1b). Pleomorphic giant cells and necrosis were absent. Mitotic figures ranged from 2-3/10 high –powered field. Metaplastic bone formation was noted at one place. The mononuclear cells expressed CD68(fig.1b,inset),tartrate-resistant acid phospatase, and smooth muscle actin, but lacked CD45,S-100 protein, desmin, and lysozyme an immunophenptypic profile identical to that of giant cell tumour of bone. Considering all these findings a final diagnosis of soft tissue giant cell tumour of low malignant potential was made.

DISCUSSION

Giant cell tumour of soft tissue(GCT-ST) is a rare tumour first described in 1972 by Salm and Sissons, followed shortly by Guccion and Enzinger [1]. Previously this tumour has been considerd to be synonymous with the giant cell variant of malignant fibrous histiocytoma with frequent local recurrence and metastasis. Recently GCT-ST has been described as a distinct entity of relatively benign prognosis, yet lacking marked atypia and pleomorphism, even in the presence of mitotic activity and vascular invasion. Some reports documented these pathological new findings, but clinical case reports are few. Biologic heterogenecity was pointed out by authors Folpe *et al* [2] who proposed a reclassification of giant cell tumour into low grade and high grade 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. Present case belong to soft tissue giant cell tumour giant cell tumour of low malignant potential [3]. Most cases of this rare tumour affects adults and the elderly and is usually located in the extremities. either superficial or deep soft tissue. Only two such cases has been reported in post. Mediastinum [4] which proves the rareity of this case. Folpe AL have studied 19 such cases in whom recurrence was seen in 4, but none developed metastasis .This contrasts with the high grade behaviour traditionally associated with MGCT of soft parts. Mononulear cells expressed CD68 and tartarate resistant acid phosphatase positivity indicating its histiocytic origin [5]. However, they lack CD45, S-100 protein, desmin, and lysozyme. For this reason this tumour has been considered the soft tissue analog of giant cell tumour of bone because of their histological and immunohistochemical similarity. This study highlights the rare location of GCT-ST of low malignant potential and emphasizes the fact that complete exicision follows a benign course because episodes of distant metastasis and tumour associated death seem to be exceedingly rare.

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