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RESEARCH ARTICLE

MEDIASTINAL TUMOURS: A CASE SERIES OF RARE TUMOURS.

Ayesha Afreen Islam[1]*, Chhanda Datta[1], Uttara Chatterjee[1], Pamela Nayak[2]

1. Dept. Of Pathology, IPGMER& SSKM HOSPITAL. KOLKATA, WEST BENGAL
2. Dept of pathology, Midnapur Medical College and Hospital, WEST BENGAL

Corresponding author: Dr. Ayesha Afreen Islam. 36 A Dr. SudhirBasu Road, Kolkata 700023. West Bengal.

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ABSTRACT:

Mediastinum, though a small anatomic compartment, diverse non-neoplastic and neoplastic pathological processes may involve it. Mediastinal masses result from a wide variety of disease processes including neoplasms, inflammatory disorders, congenital disorders, occupational disorders and autoimmune disorders. Statistically, it is important to remember that most masses (> 70%) are thymomas, lymphomas, neurogenic tumours, benign cysts and lymphadenopathy; in particular in children the most common (> 80%) are neurogenic tumours, germ cell tumours and foregut cysts while in adults the most common are thymomas, lymphomas, lymphadenopathy and thyroid masses. Here we report five very rare Mediastinal neoplasms of soft tissue origin and correlate their clinical manifestations with the histological diagnosis. Three out of our five cases were seen in young adults with age in between 17 to 20 years. All the five cases were symptomatic. Preoperative fine needle aspiration cytology was performed in four cases. Final diagnosis was achieved with histopathology. Immunohistochemistry was performed as required.

Key words: cytology, Mediastinum, thymolipoma, soft tissue giant cell tumour of low malignant potential, myoepithelial carcinoma, well differentiated liposarcoma, Lipoblastoma.

INTRODUCTION

Mediastinum is the area in the chest formed by the Mediastinal pleura laterally, sternum anteriorly and the spine posteriorly. (1) Superior border is the thoracic inlet and inferior border is the diaphragm. Mediastinal masses are a commonly encountered problem in the clinic. (1) Even though majority of these are caused by benign conditions, many of them asymptomatic, a significant proportion of these can be caused by malignant aetiology. (2)An interesting and clinically relevant feature of the Mediastinal tumours is that many of them are amenable to treatment including curative chemotherapy. (3) Despite being common, the diagnosis in an individual case can be challenging. In addition to common malignancies e.g. lymphomas and thymomas many other benign and malignant conditions can present with Mediastinal masses. (2,3) In this paper we present five cases of

Mediastinal mass and our aim is to highlight some unusual entities that present as Mediastinal masses.

CASE SERIES

Case 1: A seventeen year female was having persistent headache and swelling of face for three months before admission with episode of fever and vomiting. Physical examination revealed anaemia, facial puffiness, engorged veins in neck and upper extremity. C.T. showed a Mediastinal space occupying lesion in upper hemi thorax.(Fig 1) FNAC was inconclusive. After proper preoperative preparation and routine investigations, a plan of thoracotomy and excision of tumour was done. 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. Sections were prepared from different portions of excised mass and routine H&E stains were done followed by immunohistochemistry.

Microscopically, the tumour was composed of sheets of mononuclear cells that blended with spindled cells and benign osteoclastic giant cells (Fig 2). Pleomorphic giant cells and necrosis were absent. Mitotic figures ranged from 2-3/10 high power field. Metaplastic bone formation was noted at one place. The mononuclear cells expressed CD68, tartrate-resistant acid phosphatase, and smooth muscle actin, but lacked CD45, S-100 protein, desmin, and lysozyme. An immunophenotypic profile was 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.

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.

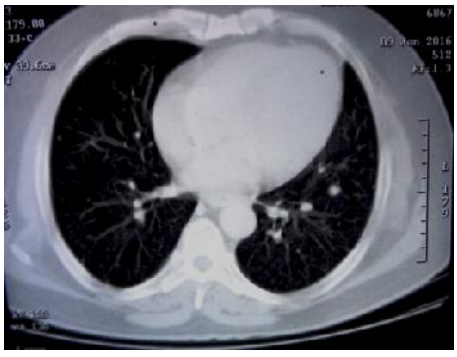

Fig 1: CT showing mass in the hemi thorax.

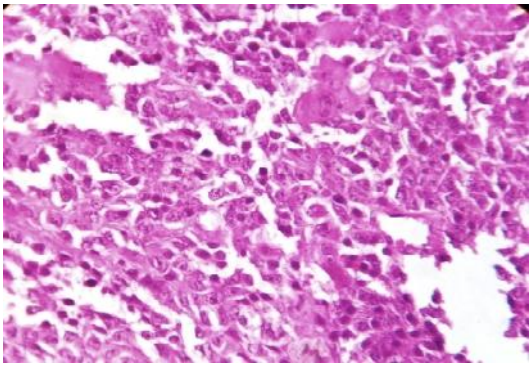

Fig 2: Pleomorphic mononuclear cells along Multi nucleated giant cells.(HPE)

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Case 2: 38 years old male presented with features of myasthenia gravis. C.T Scan chest revealed an anterior Mediastinal mass. The mass was excised and sent to our department for histopathological examination. (Fig 3)

On gross examination, there was an encapsulated yellowish mass measuring 7X7X4 cms.

Microscopic examination revealed lobules of fatty tissue admixed with normal thymic tissue. Capsular invasion noted. (Fig 4) The diagnosis of **atypical thymolipoma** was made.

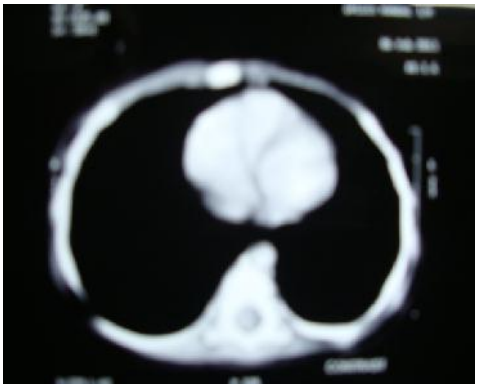

Fig 3: Anterior Mediastinal mass on CT.

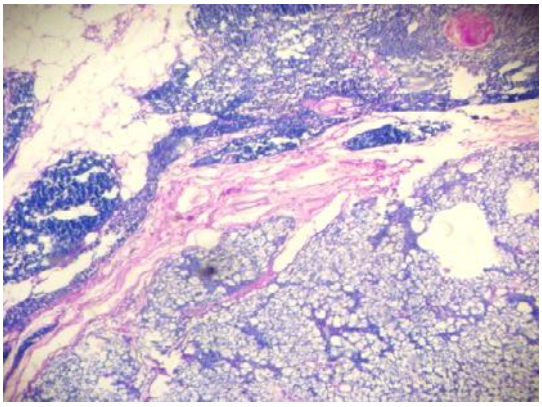

Fig 4: Pleomorphic, hyper chromatic small round Cells admixed with adipocytes and thymic tissue.

Case 3: A 19 years old female presented with features of superior vena caval syndrome and chest pain. Clinical examination revealed right sided chest dullness on percussion. Chest X-Ray and CT scan thorax showed a Mediastinal mass adherent to right lung with right sided pleural effusion and pleural thickening. Serum alfa-feto protein and beta-human chorionic gonadotropin level were within normal limit. Pleural fluid cytology was negative for malignancy. Pleural fluid ADA was 9.9 U/L and LDH was 419 IU/L. Guided biopsy from the Mediastinal mass was taken and sent for histopathological examination.

On microscopic examination, there was non encapsulated mass invading the adjacent normal lung tissue. The mass was composed of sheets of round to oval cells with clear to eosinophilic cytoplasm. The cells were positive for calponin and smooth muscle actin. A diagnosis of **Myoepithelial carcinoma** was made.

Case 4: 20 years old female presented with superior vena caval syndrome. Chest X-Ray and CT scan revealed left anterior Mediastinal mass adherent to pericardium, left phrenic nerve and left hilar structures. Guided biopsy from the mass was taken and sent to our department for histopathological examination. On microscopic examination, there was

lobules of fatty tissue composed of fat cells of different sizes admixed with scattered atypical cells. Floret cells were also noted. Atypical mitotic figures were present. Lipoblasts were not seen. The diagnosis of **atypical lipomatous tumour/well differentiated liposarcoma** was made.

Case 5: A 12-months-old boy presented with dyspnoea, stridor, and lethargy for 1 month. Computed tomography (CT) scan showed a well-defined large Mediastinal mass with density of adipose tissue (Fig 5). The mass was resected. Histopathological examination showed lobules of adipocytes separated by myxoid fibro vascular septa (Fig 6). The myxoid areas were more prominent adjacent to the fibrous septa. Adipocytes varied in degree of maturation from stellate mesenchymal cells to Lipoblasts to more mature adipocytes. The central areas were composed of mature adipocytes. Necrosis was lacking. The case was diagnosed as **Lipoblastoma**.

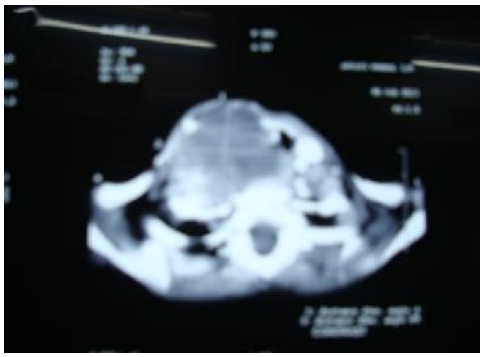

Fig 5: CT showing well defined Mediastinal mass.

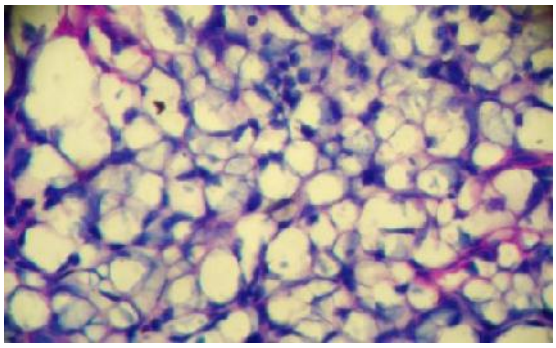

Fig 6: Varied degree of maturation of adipocytes seen on HPE.

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.(3) Previously this tumour has been considered to be synonymous with the giant cell variant of malignant fibrous histiocytoma with frequent local recurrence and metastasis.(16) 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.(16) Some reports documented these pathological new findings, but clinical case reports are few. Biologic heterogeneity was pointed out by authors Folpe et al who proposed a reclassification of giant cell tumour into low grade and high grade dependent upon the location, size, and microscopic appearance. (4,16) 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. (16) Present case belongs to soft tissue giant cell tumour giant cell tumour of low malignant potential. Most cases of this rare tumour affects adults and the

elderly and is usually located in the extremities either superficial or deep soft tissue. (10) Only two such cases have been reported in posterior Mediastinum which proves the rarity of this case. Folpe et al have studied 19 such cases in which recurrence was seen in 4, but none developed metastasis. This contrasts with the high grade behaviour traditionally associated with malignant giant cell tumour of soft parts. Mononuclear cells expressed CD68 and tartarate resistant acid phosphatase positivity indicating its histiocytic origin.(10) However, they lack CD45, S-100 protein, desmin, and lysozyme.(10) For this reason this tumour has been considered the soft tissue analogue of giant cell tumour of bone because of their histological and immuno histochemical similarity. This study highlights the rare location of GCT-ST of low malignant potential and emphasizes the fact that completes excision follows a benign course because episodes of distant metastasis and tumour associated death seem to be exceedingly rare.

Thymolipoma, a benign anterior Mediastinal neoplasm of thymic origin(5) was first described by Lange in 1916 and the term "Thymolipoma" was coined by Hall.(5) Thymolipoma accounts for less than 10 % of thymic neoplasm which itself accounts for less than 1% of all neoplasm (6). Thymolipoma usually diagnosed incidentally but less frequently associated with autoimmune diseases among them myasthenia gravis is common.(5, 8) Thymolipoma involves patients of wide age range but thymolipoma with myasthenia gravis (MG) usually seen in older individual than thymolipoma without MG.(7,9)) According to different case series, the association of myasthenia gravis with thymolipoma ranges between less than 10 to 50 percent cases of thymolipoma.(6, 8, 10) Symptoms usually relieves after surgical resection with few exceptions.(9) Pathogenesis of thymolipoma is still unclear but there were different hypothesis like this is a tumour of thymic adipose tissue, it develops as a result of replacement of thymic tissue by adipose tissue etc.(12,14) our patient was middle aged male and presented with features of myasthenia gravis. We report this case because of its rarity.

Myoepithelial carcinoma is a malignant tumour predominantly composed of myoepithelial cells and soft tissue myoepithelial carcinoma is considered as intermediate(rarely metastasizing) tumour of uncertain differentiation.(16) Myoepithelial carcinoma of soft tissue involving Mediastinum is extremely rare. Gleason BC et al. mentioned 3 Mediastinal myoepithelial carcinomas among 29 paediatric soft tissue myoepithelial carcinoma cases.(14) Papafragkou S et al. reported a single case of Mediastinal myoepithelial carcinoma in an adult female.(17) myoepithelial carcinoma show wide morphological spectrum epithelioid to spindled with eosinophilic to clear cytoplasm.(16) In Gleason's case series epithelioid morphology was predominating in majority of the cases.(17) In our case the tumour shows epithelioid morphology with eosinophilic to clear cytoplasm invading the adjacent lung. Majority of myoepithelial carcinoma cases show positivity for S100 and calponin with subset of cases show positivity for smooth muscle actin. Myoepithelial carcinoma cases show aggressive clinical behaviour with frequent recurrence and metastasis. (17, 18)

Atypical lipomatous tumour is locally aggressive tumours showing entirely or partly adipocytes proliferation of cells with significant size variation and nuclear atypia. Lipoblasts are present scattered in varying number (from many to none). Most common location is deep soft tissue of the extremities followed by retro peritoneum and groin.

Other sites accounts for less than 1% cases.(19) Chen M et al reported 10 cases of Mediastinal liposarcoma among 23 cases of primary intra thoracic liposarcoma and 4 of them were well differentiated liposarcoma in a study conducted over 10 years.(20) Boland JM et al. reported 24 cases of Mediastinal liposarcoma identified over 60 years period and 8 out of 24 cases were well differentiated liposarcoma.(21) Hahn HP et al. also reported 10 Mediastinal well differentiated liposarcoma in their case series.(21) Well differentiated liposarcoma of Mediastinum usually show better prognosis than other pathologic variant (22,23) but Matsubara H et al reported one case of Mediastinal well differentiated liposarcoma which recurs after 20 years.(14) Death occurs in 1 out of 6 cases in which follow up was possible in Boland JM case series.(21)

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