Abstract:
Angiomyolipoma (AML), the most common mesenchymal tumor of kidney, has been reported only rarely in extrarenal sites, among which liver is the commonest organ involved. Only few cases have been reported in mediastinum mostly discovered as incidental findings during investigation for other diseases. Except for liver, in all other extrarenal site AMLs, the association with tuberous sclerosis is negligible. We report two cases of mediastinal angiomyolipomas with relevant clinical details, radiologic and pathologic findings.

Keyword:
Mediastinum, Angiomyolipoma, Incidental finding

INTRODUCTION:
Angiomyolipoma is a benign neoplasm composed of abnormally thick-walled blood vessels, adipocytes and spindle & epitheloid smooth muscle cells in a variable proportion. It is generally a solitary, non-invasive tumor that most often arise from kidney. Liver is the commonest site for extrarenal AML.\(^{(1)}\)

The other extrarenal sites of occurrence include oral cavity, lymphnode, palate, spleen, vagina, spermatic cord, uterus and mediastinum\(^{(2,3)}\). About ten cases of mediastinal angiomyolipomas have been reported in the literature, so far. We report two cases of this rare neoplasm diagnosed in this year, from our department.

CASE HISTORY:
Case No.1
A 30 year old male was found incidentally to have an anterior mediastinal mass during a routine screening evaluation. CT scan showed a focal fat density lesion with hypodense areas and calcification in the paracardiac and supradiaphragmatic region with a probable radiological diagnosis of liposarcoma / thymic mass. Per-operatively a nonencapsulated mass in the anterior mediastinum anterior to left phrenic nerve was found and was seen adherent to pericardium and continuous with mediastinal fat and thymus.
Case No.2
A 57 year old male was found incidentally to have a posterior mediastinal mass during investigation for renal calculus. CT chest showed a posterior medistinal mass with central necrosis and calcification (Fig.1).

Fig.1 – CT Thorax

We received a globoid yellow-tan, capsulated masses, one measuring 8x7x4cm (Fig.2) and the other measuring 9x7x6cm (Fig.3) C/S: of both the masses showed solid, gray-white areas, yellowish fatty areas admixed with reddish-brown cystic areas.

FIG. 2A FIG. 2B

Microscopy: FIG. 4 FIG. 5

FIG. 3A FIG. 3B

PATHOLOGY: Gross
Showed a neoplasm composed of lobules of mature adipocytes and dysmorphic (thick walled and arterial like) blood vessels surrounded by bundles of spindle shaped cells. (Fig.4 & 5).

Fig.6 – Van-Gieson Stain Fig.7 – SMA Fig.8 – S100
Spindle shaped cells stained yellow with Van-Gieson stain (Fig.6). Immunomarkers were done. Spindle cells were positive for Smooth Muscle Actin (Fig.7) & Vimentin. Adipocytes were positive for S100 protein (Fig.8). With the above features a diagnosis of angiomyolipoma was made.

DISCUSSION:
Mesenchymal tumours of the mediastinum are predominantly neural in origin and are seen mostly in posterior mediastinum. Thymic tumors are commonly epithelial in nature and the only mixed mesenchymal tumors reported to arise from thymus is lipofibroadenoma (4). Angiomyolipomas of thymus / mediastinum are rare and totally only about ten cases have been reported in literature. It has been reported in 4th to 6th decade with an equal sex incidence. Though renal and hepatic angiomyolipomas show more association with tuberous sclerosis, no such association has been found in mediastinal angiomyolipomas except in one case (5). A retrospective analysis in our cases did not reveal any evidence of tuberous sclerosis. Clinically AML are usually asymptomatic and are diagnosed incidentally. Rarely may present with symptoms of tracheobronchial compression and involvement of thoracic spinal nerves. They grow insidiously and attain large size. They are seen equally in posterior and anterior mediastinum. Mediastinal AML originates from the primitive pleuripotent cells which exists anywhere in the mediastinum or spinal
cord (6) Generally it is thought to arise from Perivascular Epitheloid Cells (PEC). They are usually well encapsulated with well defined borders. However rare cases with involvement of lymphnode or extension into adjacent organs, nerves and vessels have been documented (5).

Grossly AML are encapsulated with yellowish-grey to brown cut surface and soft to elastic consistency. Microscopically, AMLs are composed of dysmorphic blood vessels with absence of elastica and lobules of adipocytes and bundles of smooth muscle cells in variable proportions. These bundles are predominantly located in perivascular region (7,8) may show ‘hair-on-end’ appearance. Smooth muscle cells may be rarely round or epitheloid in shape. Focal atypia may be seen and does not warrant a diagnosis of malignancy. More diffuse and uniform atypia and atypical mitotic figures are necessary for a diagnosis of malignant transformation in AML. Malignant transformation has been reported in renal AMLs, but not in mediastinal AMLs so far.

IHC:
The spindle cells are positive for CD117, Actin (SMA & MSA) & Desmin (9). Focal positivity for HMB-45 is seen in 85-100% of cases. Melan-A and Microphthalmia transcription factor positivity has been reported and also S100 protein, CD68 and ER/PR positivity in these tumors. They are negative for all epithelial markers. The prognosis is good. Small asymptomatic tumor may be treated conservatively with follow up. When the tumor is large sized and when there is a high risk for spontaneous rupture and hemorrhage or compression of adjacent vital structures surgical intervention is needed. Selective cases have been treated with arterial embolisation with complete cure (10).

CONCLUSION:
Angiomyolipomas are easy to diagnose as they have unique histological features. But diagnostic difficulties may arise when they are located in uncommon sites as in this case. Surgical excision is curative and it is necessary for larger lesions and the small, asymptomatic tumors can be followed up by radiological means.

Reference:

