MULTILOCULAR THYMIC CYST FROM ECTOPIC SUPERIOR MEDIASTINAL THYMIC TISSUES: A RARE CASE REPORT
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ABSTRACT: The incidence of thymic cystic lesion is rare and its occurrence from ectopic thymic tissue is rarer. We report a case of multilocular thymic cyst from an ectopic superior mediastinal thymic tissue. This 24 years young male had diagnosis of a mediastinal mass and was referred to our institute for further management. A contrast computed tomography study of chest showed a multicystic superior mediastinal mass behind the superior vanacava and innominate artery. The surgical approach was midline sternotomy and a large 20x15x20 cm mass was excised. After one year of followup he has been doing well without fresh complaints.

KEYWORDS: Mediastinum, Superior vanacava, Thymus.

INTRODUCTION: Thymic cystic lesion of mediastinum is rare and its occurrence from ectopic thymic tissue is rarer. Thymic cystic lesion are the benign lesion and can be congenital or acquired. The clinical symptoms are nonspecific and mostly due to compression to the adjacent structures. The lesion is suspected in crossectional imaging and confirmatory diagnosis is made on histopathology examination. There are some incidence of recurrence of multilocular thymic cyst after excision. These cysts are associated with thymic neoplasms such as thymoma or thymic carcinoma, and may adhere to adjacent structures and simulate as an invasive neoplasm at operation.

THE CASE: This 24 years old man presented with complaints of recurrent nonproductive cough for six months duration and on routine chest X-ray he was found to have a large homogeneous mass lesion in upper mediastinum by local treating physician (Figure 1). Later he was referred to our institute for further management. A contrast computed tomography study of chest showed a large 20x15 cm cystic mass with well-defined wall in the superior mediastinum behind the superior vanacava and innominate artery, and right of trachea and aorta (Figure 2A and 2B). The cyst showed well defined walls, multiple internal septa, and enhancing soft-tissue attenuation components. He was taken up for surgery and a median sternotomy approach was selected based on the CT scan finding of anterior relation of the mass to the great vessles. Intraoperatively the thymic tissue was found to be absent from normal anatomic location and the mass was found to be lying behind the innominate artery and superior vanacava (Figure 3). The innominate vein and superior vanacava was found to be compressed and latter was deviated towards right. The lesion was large, the size of which was nearly 20x15x20 cm and extending upto anterior paravertebral tissue posteriorly. The lesion was extracted by doing sharp and blunt finger dissection. The lesion had to be punctured to downsize it to retrieve it through limited vascular window. A histopathology examination showed multilocular cyst separated by
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fibrocollagenous septa. The cyst wall showed reactive fibrous tissue with small foci of remnant thymic tissue indicating multilocular thymic cyst. The patient’s postoperative course was uneventful and after one year of followup he has been doing well without fresh complaints.

DISCUSSION: The incidence of mediastinal cyst among mediastinal masses varies from 10 to 31%. Among the mediastinal cysts, the thymic cyst incidence varies from 3 to 13%. Thymic cysts are of two types, congenital and acquired type, with distinguished histopathology and clinical spectrum. Most of the thymic cysts are congenital. A Congenital cyst is unilocular, thin walled sac containing clear fluid, translucent and there is no evidence of inflammation. In contrast, acquired thymic cysts are multilocular and results from inflammatory process. The wall of multilocular cyst is thickened and contains thick turbid fluid or gelatinous material with features of fibrosis. Moreover, a multilocular thymic cysts may recur after excision, they may be associated with thymoma or thymic carcinoma, and they may adhere to adjacent structures simulating an invasive neoplasm at operation. There are some report of association of multilocular thymic cysts in patients with Sjögren’s syndrome, aplastic anemia, and myasthenia gravis, suggesting the possibility of an immune-mediated inflammatory process. There are some reports of implication of HIV infection, radiation, or surgical trauma in etiopathogenesis of this cyst. Cystic degeneration of a thymoma and in nearly half the patients with nodular sclerosing Hodgkin’s disease or seminoma of the anterior mediastinum have cystic thymic lesions that are histopathologically identical to idiopathic multilocular thymic cysts. Thus, in cases of suspected multilocular thymic cyst, it is of vital importance that the histology specimen be carefully examined to exclude coexisting neoplasia.

The usual clinical presentation is nonspecific and mostly due to compression to the adjacent structures like trachea or esophagus. A congenital cyst presents earlier than the usual presentation of an acquired cyst at young adult. These masses have similar imaging appearances, whatever may be its etiology. A routine chest radiogram is the primary step in evaluation and usually demonstrates it as a sharply marginated, round or oval area of increased opacity. An ultrasound may aid in the diagnosis. Contrast enhanced computed tomography shows a well-defined, heterogeneous, unilocular, or multilocular cystic masses of the thymus. The cyst wall of congenital thymic cysts is typically very thin and difficult to identify. Thus, the presence of a distinct cyst wall or the presence of calcification within the wall, indicates the diagnosis of multilocular thymic cyst. The radiologic differential diagnosis includes cystic teratoma, lymphangioma, cystic degeneration of seminoma, Hodgkin’s disease, and thymoma. Identification of fat, cartilaginous, or toothlike calcification in the lesion suggests teratoma. An alternative diagnosis should be suspected in presence of lymphadenopathy, pleural metastasis. Invasive features on CT suggest malignancy. A malignant germ cell tumor is suspected in presence of elevated serum levels of b-human chorionic gonadotropin or a-fetoprotein.

Once diagnosed, the treatment of choice is surgery. No local recurrence is reported. There has been no report of malignant degeneration of congenital thymic cyst. The prognosis is excellent after surgical resection.
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CONCLUSION: Thymic multilocular cysts are rare acquired lesions of the thymus. Its occurrence from ectopic thymic tissue is rarer. The usual etiology is inflammation but may also be associated with neoplasia. These cysts have well-defined walls, and frequently have prominent soft-tissue attenuation components. Complete surgical resection is curative and careful histopathologic examination is recommended in all patients as a CT scan cannot reliably distinguish it from neoplastic condition.

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Fig. 1: Chest X-ray shows a large homogeneous mass lesion in upper mediastinum
Fig. 2: A contrast computed tomography study of chest showed a large multicystic mass lesion in the superior mediastinum behind the superior vena cava and innominate artery.

Fig. 3: Intraoperatively a large mass was found to be lying behind the innominate artery and superior vena cava which is compressed deviated towards right.
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