CASE REPORT

Creeping mediastinal lymphangioma transgressing anatomical compartments: A rare case in an adult female patient

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Abstract

Lymphangiomas, also known as lymphatic malformations, are rare non-neoplastic lesions of vascular origin showing lymphatic differentiation. These are most commonly reported in children within the neck and axillary region; however, mediastinum remains the commonest site in adults whereby diagnosis is usually incidental on imaging done for non-specific symptoms. Radiologically, these lesions are well-defined multicystic non-enhancing masses, with CT attenuation values ranging from simple to complex fluid and fat. Being benign, these mostly present clinically either due to mass effect exerted on structures, secondarily infected or developing intra lesion haemorrhage. We present a rare case of mediastinal lymphangioma with secondary hilar and intrapulmonary extension in a middle-aged female presenting with occasional haemoptysis and shortness of breath. The patient underwent thoracotomy with complete dissection of the mediastinal tumour, per operative Bleomycin administration in pulmonary component, and made subsequent uneventful post-operative recovery.

Keywords: Mediastinal masses, Lymphatic malformations, Lymphangioma.

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Introduction

Lymphangiomas occurring within the thoracic cavity are most commonly seated within the anterior or superior mediastinum with posterior mediastinum being a rare anatomical site, statistically reported in 10% of the cases.[1] Posterior mediastinal lymphangiomas usually occur in conjunction with subdiaphragmatic extension or associated with vanishing bone disease/Gorham's disease.[1] Hilar lymphangiomas are an exceedingly rare subtype of intrathoracic lymphangiomas being reported only in a single case in isolation whilst intrapulmonary lymphangioma, described as diffuse pulmonary lymphangiomatosis, is characterised by lymphatic malformation along the bronchovascular interstitium and radiologically mimics malignant lymphangitic carcinomatosis.[2] Lymphangiomas, owing to benign proliferation of lymphatic channels, can show trans-spatial and trans-compartmental infiltration creeping and transgressing through anatomical zones with high recurrence risk and, therefore, pose significant radiological challenge in diagnosis and surgical challenge in treatment.[3,4]

Case Report

The case of a 48-year-old female, who initially came with progressively worsening shortness of breath, cough and haemoptysis for two to three years is presented. She was seen at Kalsoom International Hospital, Islamabad, on September 20, 2021. Her initial workup was unremarkable; however, upper GI endoscopy showed external compression on the oesophagus. She underwent contrast enhanced CT of the chest at the Armed Forces Institute of Radiological Imaging, Rawalpindi, in September 2021, which revealed a large non-enhancing soft tissue attenuation lesion with variable Hounsfield units centred within the posterior mediastinum causing significant mediastinal widening with mild anterior displacement of the heart. The mass draped along the anterior vertebral margin encasing the descending thoracic aorta with non-visualisation of oesophagus separately and gaining access across the perivascular route in the intra pulmonary region on the left side. No enhancing solid component, fat attenuation areas, associated mediastinal or pericardial infiltration were seen. Moreover, no transdiaphragmatic, intra-abdominal extension was noted. On the basis of imaging appearances, diagnosis of mediastinal lymphangioma with hilar pulmonary extension was suggested (Figure 1). The patient underwent video-assisted thoracic surgery (VATS) guided biopsy which suggested the diagnosis of mediastinal lymphangioma, demonstrating fibro-fatty tissue with proliferation of lymphatic channels lined by benign appearing endothelial cells. The patient underwent VATS at Kalsoom International Hospital, Islamabad, in the first week of October 2021, which demonstrated a large mass in the posterior mediastinum surrounding the aorta and oesophagus, i.e. the perihilar

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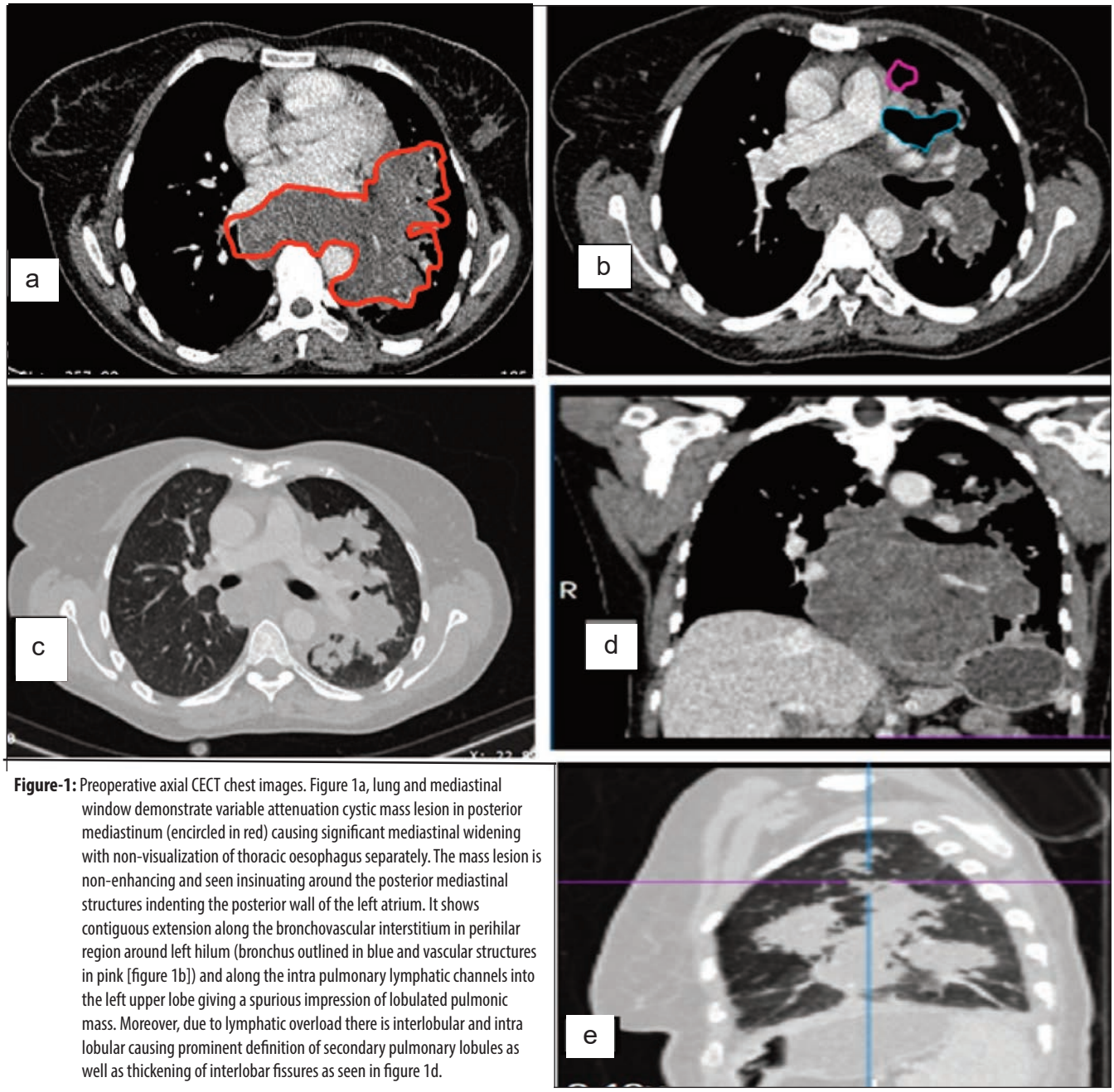

Figure-1: Preoperative axial CECT chest images. Figure 1a, lung and mediastinal window demonstrate variable attenuation cystic mass lesion in posterior mediastinum (encircled in red) causing significant mediastinal widening with non-visualization of thoracic oesophagus separately. The mass lesion is non-enhancing and seen insinuating around the posterior mediastinal structures indenting the posterior wall of the left atrium. It shows contiguous extension along the bronchovascular interstitium in perihilar region around left hilum (bronchus outlined in blue and vascular structures in pink [figure 1b]) and along the intra pulmonary lymphatic channels into the left upper lobe giving a spurious impression of lobulated pulmonic mass. Moreover, due to lymphatic overload there is interlobular and intra lobular causing prominent definition of secondary pulmonary lobules as well as thickening of interlobar fissures as seen in figure 1d.

regions, with extensions into the left lung, limited superiorly at the carina and inferiorly at the diaphragm. The mass was excised superiorly up to the level of the hilum and inferiorly to the diaphragm. The tumour was resected but pulmonary extensions into the left lower lobe and middle lobe were not excised due to their deep involvement. Bleomycin was injected into the residual mass. Postoperatively, the patient demonstrated smooth recovery. (Figure 2)

Discussion

Lymphangiomas are common mediastinal lesions in adults with posterior mediastinum being a lesser favoured anatomical location. Our case illustrates and highlights the multi-compartmental extension of lymphangioma owing to contiguous lymphatic proliferation giving constellation of mixed imaging appearances not much cited in literature.[1,2]

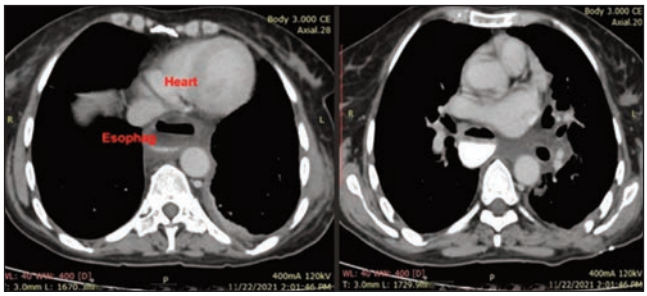

Figure-2: Post-operative follow up CECT chest mediastinal window images with positive oral contrast medium revealed non-visualisation of previously seen posterior mediastinal lesion with distended thoracic oesophagus showing air contrast level in figure 2b. There was mild mediastinal thickening (asterisk) seen at the site of previously seen lesion insinuating around the posterior mediastinal and left hilar structures in keeping with post-operative inflammatory thickening with no evidence of definite residual lesion seen.

Only 1% of all lymphangiomas are confined to the chest, comprising 0.7–4.5% of all mediastinal masses. These lesions usually have chronicity of presentation owing to the slow growing nature of the lesion and mostly remain indolent for years, presenting secondary to mass effect on adjacent mediastinal structure notably the heart and oesophagus.[2-4] Moreover, the posterior mediastinal lymphangiomas have been reported to extend sub-diaphragmatically along oesophageal/aortic diaphragmatic hiatus in abdominal cavity following the route of least resistance and subsequently being misdiagnosed on preliminary imaging as loculated abdominal ascites, omento-mesenteric or bronchogenic cyst.[4]

These usually present as cystic masses in posterior mediastinum in retro cardiac location causing significant mediastinal widening with fluid attenuation on CT. MRI can be helpful in cases where the lesion demonstrates varied density on CT and in delineation of infiltration in cardiomediastinal structures.[5,6]

Top differential considerations include thymic cyst, haematoma, seroma, or mature teratoma in case of anterior mediastinal location. The diagnosis of lymphangioma in patients with typical findings of a cystic mass in the anterior mediastinum is relatively easy. However, in atypical cases that exhibit no convincing evidence of cystic components or present in unusual locations, preoperative diagnosis may not be possible solely based on imaging characteristics.[6]

Reported recurrence rates, after complete surgical resection, are low (ranging up till 6%), whereas recurrence is reported in up to 35% in cases where resection is incomplete due to proximity to vital structures.[6,7] Pleurodesis with use of talc or other agents, including Tetracycline, Minocycline, Bleomycin and Povidone iodine, has demonstrated efficacy with success rates ranging from 80 to 100%.[7]

Conclusion

Mediastinal lymphangiomas account for 1% of all mediastinal tumours and can slowly grow to large sizes thus producing symptoms and require surgical resection. These have unique features identifiable on cross sectional imaging.

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