Incidental solitary cystic mediastinal lymphangioma with mass effect complications

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Introduction

Cystic lymphangiomas are rare benign lesions usually found during childhood in the neck, rarely extending into the superior and anterior mediastinum (Mirza et al, 2010). While intrapulmonary solitary lesions have been reported (Minato et al, 2010), solitary lesions of the mediastinum are exceedingly rare and usually asymptomatic. This article presents a case of an incidental, solitary cystic mediastinal lymphangioma with mass effect complications and the approach used in its definitive management.

Discussion

Cystic lymphangiomas, commonly named cystic hygromas (Mirza et al, 2010), are benign tumours typically located in the neck (75%) and axilla (20%) presenting before 2 years of age (Zadvinskis et al, 1992; Mirza et al, 2010). Most mediastinal lesions occur in the superior compartment as an extension of neck lesions and a small portion manifest in the anterior mediastinum (Brown et al, 1986). Fewer than 1% of cystic lymphangiomas present as solitary lesions of the mediastinum with most (90%) discovered before 2 years of age (Mirza et al, 2010; Yang et al, 2014). Although cases of cystic lymphangioma are

Case report

A 44-year-old Caucasian man with a 10 pack-year smoking history presented to the emergency department with reduced responsiveness secondary to a mixed drug overdose of fluoxetine and paracetamol. Other than a background of depression, he had no relevant past medical or family history. A plain chest X-ray revealed an incidental new mediastinal widening that was not present on a chest X-ray taken 8 years previously (Figure 1a). Thoracic computed tomography revealed a well-defined low to intermediate density lesion centred in the right paratracheal region, with associated compression of the superior vena cava, left brachiocephalic vein and displacement of the azygos (Figures 1b and c). He was referred to the 2-week wait lung cancer pathway. On further questioning he reported a 6-month history of a non-productive cough, worsening shortness of breath on exertion and chest pain radiating down the right side of the chest and shoulder. He reported losing half a stone in weight with associated lethargy. Laboratory tests were unremarkable. The case was discussed in the lung cancer multidisciplinary team meeting and referred to the cardiothoracic surgeons in view of the noted mass effect complications; he was assessed and listed for surgical resection.

Right posterolateral thoracotomy was performed through the fifth intercostal space without rib division. The large cystic lesion extended inferiorly from above the right main pulmonary artery, passing under the azygous vein and superiorly to the level of the superior vena caval origin. It was compressing the second and third intercostal nerves. A clean dissection plane allowed complete excision, and aspirate from the lesion was sent for cytology and microbiology. Enlarged lymph nodes identified at station 4 (lower paratracheal nodes) and 10 (hilar nodes) were also excised and sent for analysis.

Pathological gross examination showed a multiloculated cystic lesion 65 x 35 x 40 mm containing thin straw-coloured fluid (Figure 2a). Microscopically, it was composed of thin walls with smooth muscle and scattered clusters of lymphocytes (Figure 2b). Lining was markedly attenuated comprising bland flat endothelial cells that were positive for CD-31 and negative for AE1AE3 (Figure 2c). Pale eosinophilic fluid with lymphocytes were identified within the lesion and lymph node samples showed reactive changes.

The patient was followed up at 2 weeks and 3 months postoperatively and he was asymptomatic. He is due for a repeat thoracic computed tomography in 2 months.

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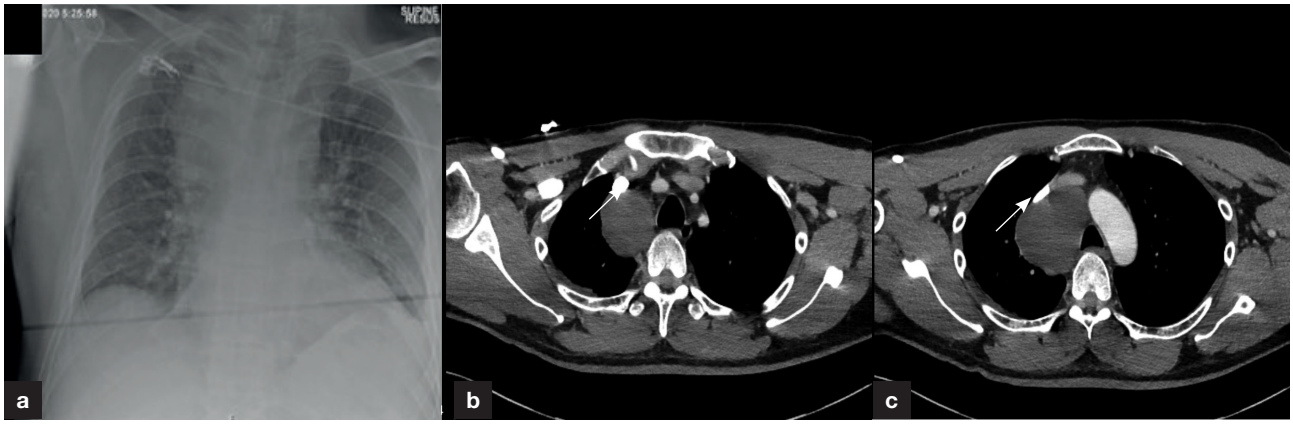

Figure 1. a. Plain chest radiograph supine anterior-posterior image shows widened mediastinum. b. Computed tomography showing a low to intermediate density mediastinal lesion (arrow) measuring 75×63 mm transaxially, (c) centred in the right paratracheal region compressing the superior vena cava.

well documented, few describe solitary cystic mediastinum lymphangiomas presenting in adulthood (Khobta et al, 2013) and none describe cases causing mass effect complications.

Lymphangiomas are congenital malformations of the lymphatic system, occurring embryologically during the development of lymphatico-venous sacs (Mirza et al, 2010). There are three types of lymphangioma: cystic, capillary and cavernous. Cystic lymphangiomas form as a result of the dilatation of inadvertently sequestered lymphatic tissue (Mirza et al, 2010). They can be several centimetres in diameter and are characterised by cystic spaces with a flat endothelial cell lining containing straw-coloured fluid with lymphocytes (Yang et al, 2014).

Although cystic mediastinum lymphangiomas are benign, growing lesions may rupture, become infected or compress neighbouring structures, such as the superior vena cava, vagus nerve and phrenic nerve (Misthos et al, 2006). Recurrences are uncommon, except in cases where the tumour has not been completely resected (Brown et al, 1986). This case

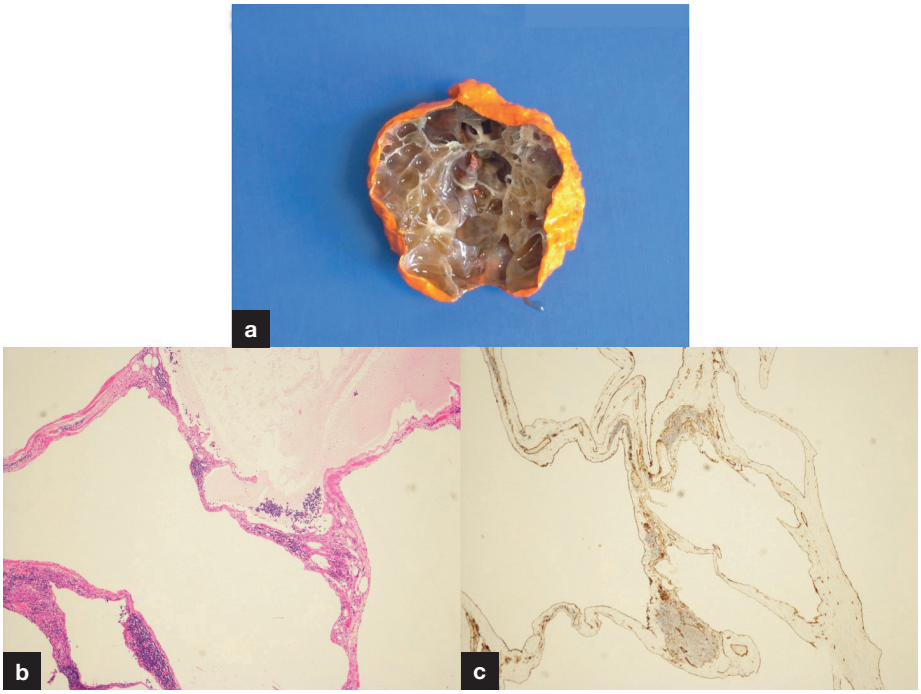

Figure 2. a. Gross image of cystic mass with a multi-loculated cut surface. b. Histological image showing thin walls containing smooth muscle and clusters of lymphocytes (haematoxylin and eosin stain ×40). c. Histological image showing a markedly attenuated lining comprising bland flat endothelial cells that are positive for CD-31 (×40).

Learning points

- Although cystic mediastinum lymphangiomas are benign, their location and growth in the mediastinum can cause compression of surrounding structures.
- Early detection and complete surgical resection are paramount to prevent complications and recurrence.
- Early cardiothoracic surgical opinion should be sought in all cases.

highlights the importance of investigating persistent chest symptoms, including chest pain and shortness of breath. In addition to a detailed clinical assessment, a plain chest X-ray should be considered to identify masses. Complete surgical resection is the treatment of choice to prevent potential mass effect complications.

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