CASE REPORT

Giant Cystic Lymphangioma of the Middle Mediastinum

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ABSTRACT

A middle-aged male presenting with complaint of progressively increasing dysphagia was found to have a large cystic mass lesion in the middle mediastinum on evaluation. A diagnosis of an endo-duplication cyst was considered after exploration, in view of infiltration of the muscular layer of the lower thoracic esophagus, presence of multiple hyperemic nodular lesions on its inner surface and its location in the middle mediastinum. However, the histopathology revealed the lesion to be a cystic lymphangioma.

Key words: Cystic lymphangioma, Middle mediastinum, Dysphagia.

INTRODUCTION

Cystic lymphangiomas are rare benign tumours of lymphatic origin\(^1\). Majority of these lesions are congenital in origin and are seen in children, presenting as a soft neck mass\(^1,2\). These cervical lesions may have an extension into the superior or anterior mediastinum\(^1,2\). Lymphangioma totally confined to the mediastinum are extremely rare and have been reported mostly in adults\(^1,2\).

Though usually asymptomatic, a large lesion extending into superior mediastinum may cause obstructive respiratory symptoms or features of venous obstruction\(^1,3\). Isolated mediastinal lymphangioma are usually diagnosed incidentally, late in life because of lack of symptoms\(^2,3\).

We present a case of a giant cystic lymphangioma of the middle mediastinum, remote from the neck, in an adult patient who presented with dysphagia.

CASE REPORT

A 43-year-old male patient presented with a symptom of gradually increasing dysphagia to solids since nine months with a significant loss of weight. Physical examination of the patient was unremarkable, except for mild anemia.

Biochemical tests revealed a low haematocrit of 27 percent. Liver function tests, alphafetoproteins and human chorio-gonadotrophins were within normal range. Chest roentgenogram (Figure 1a) showed evidence of a mass lesion in the lower part of the mediastinum, extending onto the right side of the chest and associated with collapse of the right middle

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lobe. Computed tomogram of the chest (Figure 1b) showed a large cystic mass with water density in the middle mediastinum, pushing the heart anteriorly and the esophagus posteriorly. The lesion was seen extending from behind the lower trachea to the level of the diaphragm.

**Figure 1a.** Chest roentgenogram (PA view) showing evidence of a mass lesion in the lower part of mediastinum, extending onto the right side of the chest.

**Figure 1b.** Computed tomogram of the chest showing a large cystic mass with water density in the middle mediastinum, pushing the heart anteriorly and the esophagus posteriorly.

Barium swallow (Figure 1c) revealed the esophagus to be compressed and displaced to the right but without any hold-up of the contrast or mucosal irregularity. On bronchoscopy, the right middle bronchus was found compressed with no mucosal lesion. Oesophagoscopy showed evidence of an external compression at about 24 cm. The oesophageal mucosa appeared normal. On two dimensional echocardiography, the left atrium was found to be compressed by an extraneous mass. There was no infiltration into the pericardium or evidence of pericardial effusion.

With a pre-operative diagnosis of an endoduplication cyst, the patient was taken up for exploration, through a right posterolateral thoracotomy incision and controlled lung ventilation. A tense cystic mass (15 cm x 10 cm x 7 cm in size) with bosselated surface was seen lying between the heart and the vertebral bodies, displacing the heart to extreme left, and extending from the level of tracheal bifurcation to the diaphragm. The middle lobe of the right lung was collapsed and adherent to it. The esophagus was pushed posteriorly and to the right side and the mass was infiltrating the muscular layer at its lower one-third. The cyst
was separated entirely from its surrounding structures and excised, along with the anterior esophageal muscular layer at the point of infiltration. The esophagus was then reconstructed.

The cyst was filled with straw coloured fluid. The wall of the cyst was irregularly thickened and had multiple nodular lesions on its inner surface, which was also, trabeculated. The operative diagnosis was presumed to be a reduplication cyst of the esophagus with ectopic gastric mucosa. However, on histopathological examination, the lesion was reported as cystic lymphangioma (Figure 2).

(Figure 3). The patient had completed fourteen months of follow-up, without any delayed complication,

Figure 2. Photomicrograph showing dilated lymphatic channels surrounded by aggregates of lymphocytes (H & E x 200).

Histopathological study showed the cystic wall devoid of any lining epithelium. Instead, it had a thick fibrous layer lined by granulation tissue composed of diffuse lymphoplasmacytic infiltrate, congested proliferating capillaries and plump myofibroblasts, plasma cells and haemosiderin laden macrophages. In addition, it showed dilated lymphatics with smooth muscle fibers in the wall and endothelial pfolifertaion along with peri-lymphatic lymphoplasmacytic infiltration. Few smooth muscle fibers were also seen at the periphery, in the area where the cyst was infiltrating the muscular layer of the esophagus.

Patient had smooth post-operative recovery. Chest roentgenogram showed clear lung fields with expansion of the right middle lobe. Follow-up barium swallow revealed normal esophagus

DISCUSSION

Mediastinal cystic lymphangiomas are extremely rare benign tumours. Less than one percent of cystic lymphangioma are reported to arise from the mediastinum. Cystic lymphangioma present in two clinico-pathologic forms. The more common variety of lymphangioma, called cystic hygroma, is usually seen in infants as a soft cystic swelling, either localized entirely to the neck or with extension into the superior or anterior mediastinum or the apical pleural space. The type of lymphangioma probably represents a developmental anomaly rather than a true neoplasm, resulting from failure of lymphatic system to communicate with the venous system. Isolated mediastinal lymphangioma are only infrequently seen in children. Other type of lymphangiomas are seen mostly in adults and are more or less well
circumscribed lesions in the anterior, middle or posterior mediastinum, remote from the neck\textsuperscript{14}. The origin of purely mediastinal lymphangioma is unclear. It has been suggested that these probably arise from mesenchymal deposits left in the mediastinum during embryonic development\textsuperscript{5} or represent a true neoplasm arising from the mediastinal lymphatics\textsuperscript{6}.

Due to their cystic nature and soft consistency, lymphangioma seldom cause symptoms even when large and are more often diagnosed incidentally on chest radiography\textsuperscript{7,8}. Only occasionally, a large cystic hygroma in the anterior or superior mediastinum may produce symptoms by compressing the adjacent structures, such as the trachea-bronchial tree or the superior vena cava or its major tributaries\textsuperscript{1,2}. Posterior mediastinal lymphangioma do so rarely\textsuperscript{1,9}. To our knowledge, there is no report of an isolated lesion in the middle mediastinum producing symptoms from displacement or compression of the adjoining structures. All the reported lymphangioma in this location have been discovered incidentally\textsuperscript{7,10}.

The radiographic findings of a mediastinal lymphangioma are non-specific and mostly consist of evidence of a smoothly marginated mediastinal mass. Though cystic hygroma of the neck has a characteristic sonographic appearance, mediastinal lesions are better evaluated with computed tomography scans, which show a well-defined homogeneous water density mass enveloping adjacent mediastinal structures without any contrast enhancements.

Complete excision is the only effective treatment of cystic lymphangioma. During the operation, one should develop a plane of dissection that leaves the capsule intact because, after rupture, the decompressed cyst may be difficult to handle, predisposing to incomplete removal and a risk of recurrence\textsuperscript{2}. Alternative therapies such as aspiration, incision and drainage, irradiation, or injection of sclerosing agents have all been used for the management of cervical cystic hygroma, with little success. These have no role in the treatment of mediastinal lymphangioma\textsuperscript{2}.

Our case was unusual on two counts. First, the patient was middle-aged with progressively increasing dysphagia. Such a presentation have not been previously described in association with mediastinal lymphangioma. Secondly, the tumour was found to be in the middle mediastinum, a very rare location and had attained a large size (15 cm x 10 cm x 7 cm). Due to the location of the lesion in the middle mediastinum, its size and extent from tracheal bifurcation to the diaphragm and its intimate relation with esophagus during exploration, the diagnosis of an enteral duplication cyst was considered, which is more likely to present in this manner. Cystic lymphangioma was never suspected until the histopathology report. By way of its size and location, it had produced significant compression of the esophagus causing dysphagia. Investigations were inconclusive in diagnosing the exact pathology. Exploratory thoracotomy provided the material for diagnosis, in addition to relieving the symptoms.

In conclusion, though rare, cystic lymphangioma should be considered in the differential diagnosis of cystic lesions of the mediastinum, irrespective of their location or age of the patient. Surgical excision is the treatment of choice, as it provides complete cure and symptomatic relief to the patient and allows precise diagnosis on histopathological examination.

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