

Case reports

Lymphangiomatosis presenting with bronchial cast formation

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Abstract

Lymphangiomatosis is a rare disorder of the lymphatic system that is known to cause chylothorax. Chyloptysis may occur but chylous bronchial cast formation is rare. A case is reported of lymphangiomatosis in a 34 year old woman whose initial manifestation was cough productive of bronchial casts. Two years later the patient developed a chylothorax. Ligation of the thoracic duct through a low thoracotomy was curative.

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Keywords: lymphangiomatosis, lymphangiectasis, chyloptysis, bronchial casts.

A 34 year old non-smoking woman presented in 1990 with persistent cough for one year. The cough was intermittently productive, was blood tinged, and contained fine branching bronchial casts. She was otherwise well and taking no medication. Examination and routine laboratory evaluation including eosinophil count were unremarkable. Arterial blood gas tensions breathing room air showed a pH of 7.46, PCO_2 of 4.7 kPa, and a PO_2 of 9.5 kPa. A chest radiograph revealed increased linear markings in

the middle and lower lung fields. Pulmonary function tests revealed a forced expiratory volume in one second (FEV_1) of 2.21 (71% predicted), forced vital capacity (FVC) of 2.75 l (71% predicted), FEV_1/FVC of 80%, total lung capacity (TLC) of 4.3 (77% predicted), and a transfer factor of 17.2 mmol/min/kPa (78% predicted). Sputum culture grew normal flora. A computed tomographic (CT) scan of the thorax revealed mucous impacted in the bronchi. Defects in the liver and spleen were seen consistent with cysts or haemangiomas. There was no evidence of bronchiectasis. Sweat chloride levels and quantitative immunoglobulins including IgE were normal. Aspergillus skin test was positive but aspergillus precipitins were negative.

The patient underwent fiberoptic bronchoscopy in February 1991. Many casts were aspirated that floated in normal saline which, on microscopic examination, were found to be composed of mucin, scattered inflammatory cells, and macrophages. Cultures and bronchial biopsy samples were unrevealing. A diagnosis of plastic bronchitis¹ was made and treatment with bronchodilators, mucolytics, intermittent corticosteroids, and antibiotics was instituted. Nonetheless, over the next 18 months, episodic dyspnoea, wheezing, and cough productive of bronchial casts continued.

In August 1992 she presented with three days of shortness of breath and a respiratory rate of 40 breaths per minute. Chest examination revealed dullness to percussion bilaterally with decreased breath sounds at the bases, diffuse wheezes, and coarse crackles. The white blood count was 13 500/mm³ with a normal differential. Chest radiography showed increased interstitial markings and bilateral effusions. Milky pleural fluid was withdrawn on thoracentesis. The triglyceride level was 832 mg/dl.

High resolution CT scanning showed small nodular densities in the para-aortic area and aortopulmonary window that were not present in 1990. A lymphangiogram with delayed upright views at 24 hours detected reflux of contrast into the left lung and fluid/fluid levels in the region of the thoracic duct (fig 1). The patient underwent a left anterior mediastinotomy. Numerous clusters of dilated lymphatics were seen in the mediastinum. Biopsy specimens revealed dilated proliferative lymphatic channels consistent with lymphangiomatosis (fig 2).

The patient required bilateral tube thoracostomy due to increasing effusions and underwent a thoracic duct ligation via a low right thoracotomy. The chylous drainage resolved postoperatively.

Two years later the patient is well. The pleural effusions have not reaccumulated. There is no evidence of lymphoedema and pulmonary function tests are normal.

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Figure 1 Lymphangiogram showing fluid/fluid levels (arrow) in the mediastinum.

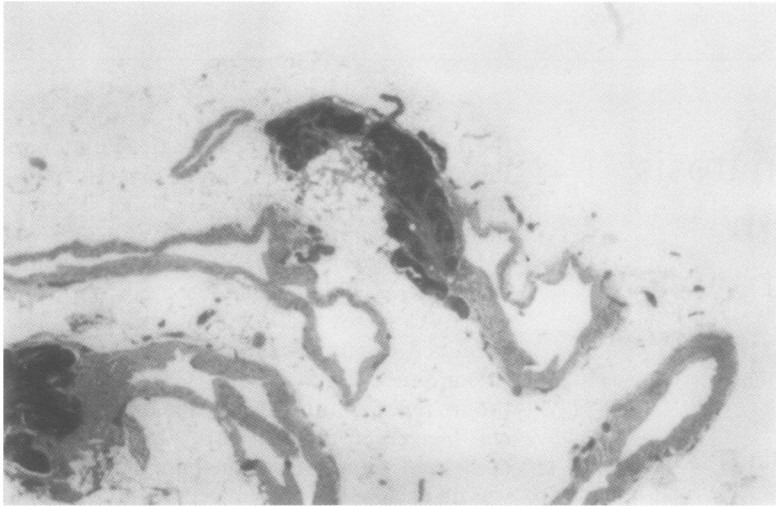


Figure 2 Mediastinal biopsy specimen showing dilated proliferative lymphatic channels consistent with lymphangiomatosis (original magnification $\times 10$, reduced to 62% in origination).

Discussion

Abnormal pulmonary lymphatic drainage can occur when there is congenital malformation of lymphatics or when flow through the thoracic duct is disrupted. Congenital malformation of lymphatic vessels is rare and has been classified into lymphangiectasis (abnormal dilatation) and lymphangiomatosis (abnormal proliferation), although both processes may be seen in the same biopsy specimen.² The lymphatic abnormality may be isolated to the thorax or be part of a generalised process.^{3,4}

Patients with primary lymphatic disorders most often present in infancy and early childhood but cases have been reported with onset of symptoms as late as the eighth decade.⁵ Findings at the time of presentation may include a discrete mass in the neck, mediastinum, or axilla. In some, chylothorax is the initial manifestation. Chyloptysis, the expectoration of milky sputum, is thought to be due to stasis of lymph flow with reflux of chyle into the tracheobronchial tree.

This patient presented with chronic expectoration of bronchial casts. Familiar causes of bronchial casts include asthma, allergic bronchopulmonary aspergillosis, cystic fibrosis,

bronchiectasis, bacterial infection, and plastic bronchitis,⁶ but chyloptysis is not routinely cited. In 1968 Maier⁷ reviewed 17 cases of "chyloous reflux into the lungs and pleurae" and found that two of six patients with chyloptysis had cast formation. Wetherill *et al* have also described a case in which chyloous bronchial casts were expectorated.⁸ The sputum in that case was confirmed to be chyloous by triglyceride analysis and the casts were noted to float in normal saline. An anatomical correlation between casts and a lymphatic abnormality was noted by Wiggins *et al*⁹ who reported a patient who expectorated casts for 23 years. The right middle lobe was eventually resected and dilated lymphatic vessels were noted in the hilum.

The diagnosis of lymphangiomatosis may be confirmed by lymphangiography with delayed upright views.¹⁰ Surgical resection may be difficult if lesions are diffuse. Other management options include observation,⁸ ligation of the thoracic duct,^{4,5} pleurodesis or parietal pleurectomy,⁷ low fat diet plus progesterone therapy,² chemotherapy, and radiation.

Of these, thoracic duct ligation may prevent complications such as malnutrition, pulmonary fibrosis, and respiratory failure.^{4,5,8} A recent review of Gorham's syndrome complicated by chylothorax⁴ cited a survival rate of 72% in patients who underwent thoracic duct ligation compared with 36% without surgery.

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