Pleuro-Pulmonary Lymphangiomatosis: Malignant Behavior Of A Benign Disease

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Citation

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Abstract

Lymphangiomatosis is a rarely encountered congenital derangement of the lymphatic system, usually presenting in late childhood that sometimes goes under-recognized. It occasionally behaves as a malignant disease and is completely different from another entity called lymphangioleiomyomatosis. We describe a case of a young patient who presented with dyspnea and was found to have massive involvement of the right pleura, pericardium, mediastinum and right axilla, mimicking mesothelioma.

ABBREVIATIONS

LAM = lymphangioleiomyomatosis
CT = Computed Tomography
SVC = Superior Vena Cava

INTRODUCTION

Lymphangiomatosis is a congenital derangement of the lymphatic system presenting in late childhood. Although histologically benign, it has propensity to infiltrate surrounding tissues. Very few cases of thoracic lymphangiomatosis have been reported in the literature. Here, we describe a case presenting with a cystic and solid pleuro- pulmonary mass arising from the right hemithorax and encasing the lung, mediastinum and axilla.

CASE REPORT

The patient was a 17 year-old Caucasian female presenting with dyspnea. Two years back she developed chest pain. She was then diagnosed with pneumonia, and given antibiotics without relief. Her chest pain became more intense and was admitted to the hospital. CT scan of the chest revealed a circumferential thickening of the right pleura encasing the lung and extending into the mediastinum, both hila , pericardium, pulmonary veins and celiac axis. There were multiple cystic components of the mass compressing the adjacent parenchyma. There was also right axillary, retropectoralis, and right anterior diaphragmatic adenopathy present (Figures 1 and 2). Subsequently, she was taken to surgery and biopsy showed the mass not to be malignant. According to the surgeon: “the mass felt like concrete”.

Figure 1

Figure 1: Pleuro-pulmonary lymphangiomatosis. Contrast-enhanced CT scan of the chest at the level of innominate veins showing a right axillary mass and small nodes, and circumferential thickening of the pleura, encasing the right lung, trachea, esophagus and major vessels.

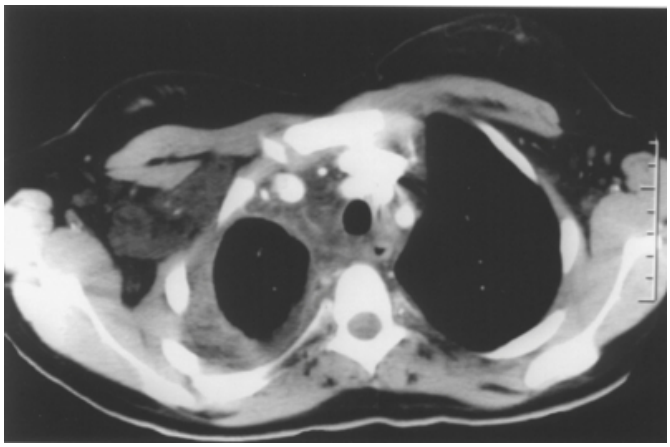
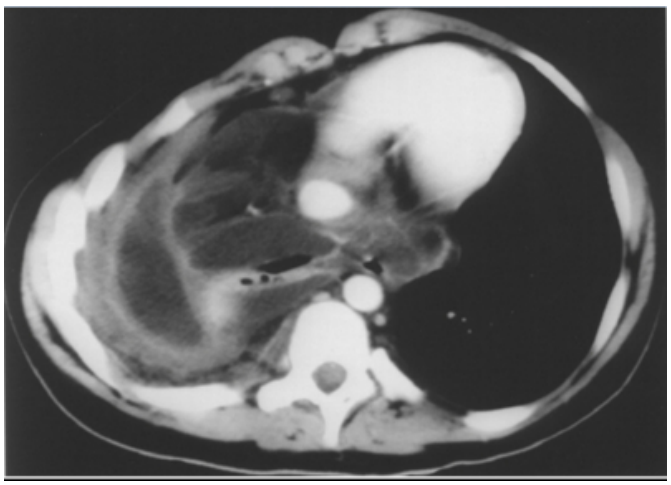


Figure 2

Figure 2: Pleuro-pulmonary lymphangiomatosis. Contrast-enhanced CT scan of the chest at the level of cardiac ventricles showing diffuse thickening of the pleura, multiple chylus lakes extending into the pericardial sac and mediastinum. Note restrictive collapse of the right lung and an enlarged right anterior diaphragmatic lymph node.



Pulmonary function tests showed decreased lung volumes, which did not change in a six month-period. CT of the chest taken one year later showed that the mass was unchanged. Patient's symptoms did not change significantly during this period.

Her past medical history was significant for 10-degree scoliosis. Patient's father and brother had ankylosing spondylitis; paternal grandfather had lung cancer.

Physical examination was remarkable for decreased breath sounds and crackles on the right lung base. A small, nontender nodule was palpable in the right axilla.

Review of the submitted pathological material was compatible with lymphangiomatosis and there were foci of hemorrhage and fibrosis.

In view of patient's stable condition and the low benefit/risk ratio of chemotherapy, treatment was withheld.

DISCUSSION

Diseases of the pulmonary lymphatic system occur in a variety of clinical settings. Errors of development can lead to primary pulmonary lymphatic disorders that, when focal, are termed lymphangiomas, and when diffuse, are named lymphangiomatosis. The latter form is frequently associated with other lymphatic abnormalities that may involve multiple organ systems.

Congenital lymphangiomatosis may be due to failure of lymphatic channels to regress after the 20^{th} fetal week, or failure to connect to efferent channels, or due to sequestration of lymph sacs during development. Acquired forms are usually the result of chronic inflammation or infection.

Lymphangiomatosis usually presents in late childhood, as in this case. It can infiltrate the adjacent tissues, and mimic malignant processes i.e., mesothelioma, however, histopathological evaluation demonstrates mature cells and dilated lymphatic channels lined by endothelium. The cystic spaces are filled with proteinaceous lymph fluid. Lymphoid aggregates are benign and cellular components are well differentiated.

The clinical manifestations (such as cough or dyspnea) develop after a period of latency. They may not become apparent until restrictive compression of vital structures develops. The presenting symptom can be dyspnea as in our case, or symptoms related to dysphagia, stridor, Horner's syndrome, superior vena cava syndrome or phrenic nerve palsy. It can also be discovered as an incidental finding on imaging studies.

Bony involvement and chylous effusions are common. Pulmonary function tests may reveal a mixed obstructive and restrictive pattern. In our case, restrictive features were predominant.

Surgical resection or sclerotherapy are the therapies of choice when the mass is localized[1, 2]. Complete surgical resection may prove technically difficult when there is diffuse involvement, because of infiltrative nature of the disease, resulting in recurrence and return of symptoms. Therapy should be aimed to decrease the symptoms that arise from compressive effects, to control chylous effusions, and to maintain optimal cosmesis. In patients with widespread disease, palliation is the goal of therapy. Low fat, high protein diet with medium chain triglycerides helps to reduce the lymph flow. Serum immunoglobulin level monitoring is recommended.

Because of name similarity, this condition could be confused with lymphangioleiomyomatosis (LAM). LAM is caused by a proliferation of abnormal smooth muscle cells around airways, blood and lymphatic vessels, resulting in obstruction and dilatation of these structures, leading to air trapping, hemoptysis, and chylothorax, respectively. In contrast to LAM, the lung parenchyma is preserved in

lymphangiomatosis[3]. LAM usually occurs in premenopausal women and causes restrictive/obstructive ventilatory defects.

The differential diagnosis should include processes with massive pleural involvement that may extend into the mediastinum such as mesothelioma, malignant fibrous histiocytoma, metastatic processes, and fibrosing mediastinitis.

CONCLUSION

Lymphangiomatoses are late childhood disorders of the lymphatic system, not commonly encountered in clinical practice and yet under-recognized. Nevertheless, considering their crippling nature, they must be acknowledged, because effective management might be facilitated by early and accurate diagnosis.

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