Case Report

A rare case of pulmonary lymphangiomatosis from the tribal zone of central India

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Received: 13 July 2015
Revised: 14 July 2015
Accepted: 11 August 2015

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ABSTRACT

Lymphangiomatosis, a rare disease, occurs in individuals of any age, regardless of gender, but is predominantly seen in younger individuals. It often presents with pulmonary involvement, although, the bones, spleen and liver can also be affected. Histologically, pulmonary involvement includes proliferation, complex anastomoses and secondary dilatation of the lymphatic vessels. Clinically presentation is often variable but pulmonary involvement is affected more common than other involvements. Diagnosis is made histologically but radiologically can suggest diseases. Treatment is only supportive and symptomatic.

Keywords: DPL (Diffuse pulmonary lymphangiomatosis), VEGFR-3 (Vascular endothelial growth factor receptor 3)

INTRODUCTION

Pulmonary lymphangiomatosis is a rare clinical disease and pathological entity characterized by proliferating lymphatic vessels, multiple lymphangiomas in the lungs liver, soft tissues, bone, spleen, mediastinum. It is controversial whether they are neoplastic or hamartomatous in origin1. The clinical course of patients with lymphangiomas is relatively progressive and benign. The disease is usually fatal in infancy and early childhood if associated with congenital anomalies.2 The onset of clinical manifestation is unusual can occur in any age group but usually occur in adult group. Diffuse lymphangiomatosis affects a variety of tissues, and the term diffuse pulmonary lymphangiomatosis is used when the alterations are restricted to the chest. Diffuse lymphangiomatosis is the most common form.3

CASE REPORT

A female patient age of 46 year admitted in Government Medical College Hospital Rajnandgaon with complain of progressive breathless on exertion and non-productive cough, chest tightness since five years and recently developed swelling over both lower limbs and distention of abdomen with loss of appetite over two weeks Patient is belongs to rural area and by occupation she is house wife and there is no addiction history to patients. Past history of patient is insignificant.

Patient was investigated her hemoglobin was 12 gram, urea was slightly elevated 49.5 mg/dl creatinine was normal other test liver function test and urine microscopy was normal. On ultra sonography abdomen hepatomegaly was seen and no other abnormality was detected. Radiologically X-ray chest there is multiple emphysemous bulla seen throughout the lung field, with
shifting of trachea to left side with thickening of pleura with blunting of both costophrenic angle with patchy infiltration in both lower lung field. On CT chest there variable cystic change seen in both lung parenchyma with pneumatocele with mediastinal shifting and pleural thickening. Two dimensional echocardiography was normal with ejection fraction of 60% with no pulmonary hypertension with all cardiac chamber of same size. Patient was managed supportive with glucocorticoids and bronchodilator and patient is now symptomatic better.

![Figure 1: Radiologically CT scan thorax.](image)

**DISCUSSION**

Pulmonary lymphangiomatosis is a rare pulmonary lymphatic system disorder involving the entire lymphatic system from the mediastinum to the pleura. Although it is pathologically benign, it is a progressive and fatal disease. Histologically observed that there is an increase in the diameter and number of involved lymphatics. The disease is generally seen among children and adolescents. Histologically, it is difficult to differentiate it from other lymphatic diseases like lymphangiomia, lymphangiomyomatosis, and lymphangiectasia. Diffuse Pulmonary lymphangiomatosis presents itself with diffuse mediastinal soft tissue infiltration, pulmonary interstitial parenchymal infiltration, and pleural effusion. Lymphangiomatosis can potentially affect any part of the body except the brain. The disorder can be widespread, affecting multiple areas simultaneously, as in the case of diffuse pulmonary lymphangiomatosis, or be isolated to one area (i.e. the lungs and chest). The specific symptoms and severity vary, depending in part upon the size and the specific location involvement. Diffuse pulmonary lymphangiomatosis causes functional impairment of the lungs and when the chest wall is involved, may be associated with disfigurement.

**Etiology**

The exact cause of pulmonary lymphangiomatosis is unknown. It is believed to result from abnormalities in the development of the lymphatic vascular system during embryonic growth. No environmental, immunological or genetic risk factors that may play a role in the development of the disorder have been identified. Different studies show that whether Vascular Endothelial Growth Factor Receptor 3 (VEGFR-3) plays a role in the development of lymphangiomatosis. Affected tissue in individuals with lymphangiomatosis have high levels of VEGFR-3, a chemical that most likely promotes the growth of lymphatic vessels and tissue of the affected body parts.

**Clinical presentation**

Diffuse pulmonary lymphangiomatosis affects males and females in equal numbers. Cases have been reported infants and children, but the disease has occurred in adults as well. The symptoms of pulmonary lymphangiomatosis are caused by complications due to the proliferation, dilation, and thickening of lymphatic vessels throughout the lungs. There is progressive development of breathless on exertion initially then at rest, with non-productive cough, hemoptysis chest pain, wheezing and chest tightness and recurrent respiratory tract infection can occur. Atypical presentation may occur if involvements of other system of body parts such as cardiac tamponade and pericardial effusion and heart failure. Disease is usually fatal and leads to respiratory failure and ultimately leads to death within decades.

Other related disease may have similar presentation these include hemangiomatosis congenital pulmonary lymphangiectasia, lymphangioleiomyomatosis, various tumors or malignancies including hemangioendothelioma and Kaposi’s sarcoma of the lung. Several different obstructive lung diseases pulmonary, such disorders include, emphysema, asthma chronic obstructive bronchitis, and interstitial lung disease.

**Diagnosis**

Diagnosis is usually made definitive by histology of affected parts but other radiological tools can help for differentiating from other disorder. A diagnosis of diffuse pulmonary lymphangiomatosis is based upon identification of characteristic symptoms, a detailed patient history, a thorough clinical evaluation, and a variety of tests. Because disease is rare varying presentation in each individual, obtaining a correct diagnosis can be very difficult.

**Workup**

Various imaging techniques including plain X-rays, ultrasound, Computedized Tomography (CT) scanning and Magnetic Resonance Imaging (MRI) may be used to determine the location, behavior and extent of the disorder.
Chest radiographs are nonspecific and can reveal diffuse interstitial infiltrates and pleural effusions and pleural thickening and infiltrates in lung parenchyma. Chest CT are more specific include peribronchovascular and interlobular septal thickening, diffuse liquid-like infiltration of the mediastinal soft tissue and pleural thickening with effusions. These findings are suggestive - but are not pathognomonic can occur in other disease also. Any part of the mediastinum can be affected - anterior, middle, posterior, and superior, perhaps with slight prevalence in the anterior part and there is well defined multilocular water density mass with or without compression or displacement of nearby structures.9-11

Pulmonary function tests may be used to help diagnose diffuse pulmonary lymphangiomatosis. Affected individuals often have a restrictive pattern or a mixed restrictive and obstructive pattern. Echocardiography may be helpful and more extensive provide detailed descriptions and allow for an accurate diagnosis. Furthermore, MRI can be a useful method to achieve a differential diagnosis. However, conventional coronary angiography can visualize tumor vascularity, but the modality is not considered a primary imaging method.12

Lymphoscintigraphy is a specialized procedure in which small amounts of radioactive material is used to help create pictures (called scintigrams) of the lymphatic system. Lymphoscintigraphy is used to help obtain a diagnosis of lymphatic disease and to assess the extent of the disease.13

Surgical removal and histological examination of affected lung tissue (lung biopsy) may be used to confirm a diagnosis of diffuse pulmonary lymphangiomatosis. However, a lung biopsy is not always possible and can be associated with complications. Bronchoscopy is nonspecific and can reveal airway mucosal erythema and edema, bronchial narrowing and, in advanced cases, thin-walled vesicles containing chylous fluid.14,15 Differential diagnosis include lymphangiomatosis, lymphangiectasis, lymphangiomatosis and lymphangiomatosis. These have several shared pathological characteristics which lead to difficulties in differential diagnosis.16

Treatment

Prognosis is generally poor and treatment is mostly symptomatic because there are no definitive treatments available for this disease. Surgical resection for localized lesion or mediastinal lesions, thoracotomy has been reported to be effective.17 Other attempted include thoracocentesis, parietal pleurectomy and ligation of the thoracic duct. However, with the diffuse type when there is multiorgan involvement i.e., heart, pleura, pericardium, ribs, and thoracic vertebrae, the role of surgery is limited apart from a biopsy for tissue diagnosis or draining of pericardial effusions and aspiration with pleurodesis for patients with recurrent pleural effusion. Low-fat in diet with medium-chain triglycerides have been attempted but are minimally successful. Therapeutic thoracocentesis and pleurodesis for recurrent pleural effusions for symptomatic relief.17

Systemic glucocorticoids, recombinant interferon therapy, cyclophosphamide, tamoxifen and radiation therapy may decrease parenchymal involvement and incidence of effusion. These treatment have less success rate but more toxicity.16,17

Treatment with propranolol, sirolimus and bevacizumab has been shown to decrease recurrent pleural effusions and lymphatic proliferation with less severe toxicity are the recent treatment modalities. Other treatment options include palliative surgical treatment or the use of sclerosing agents, such as OK-432 and palliative irradiation can be useful. Lung transplantation can be one of the treatment modalities.18

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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