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CASE REPORT

LUNG CYSTS IN CLINICAL PRACTICE; A CASE OF SPORADIC LYMPHANGIOLEIOMYOMATOSIS IN A POST MENOPAUSAL LADY

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ABSTRACT

Lymphangioleiomyomatosis (LAM) is a form of cystic lung disease, quite rare disease of unknown cause that traditionally affects young women of reproductive age. It is characterized by excessive proliferation of immature smooth muscle and spindle cells (LAM cell) along the axial lymphatics, leading to progressive cystic lung destruction, lymphatic obstruction, and abdominal tumours. LAM is almost universally fatal without a lung transplant. Because of its rareness and nonspecific presenting symptoms, patients often receive a missed or delayed diagnosis. We present the case of a 68-year-old postmenopausal woman from maldives who presented with fever, cough with mucopulent phlegm and worsening orthopnoea was ultimately determined to be due to LAM with Acenetobactor infection. LAM may be more commonly recognised than previously and is imperative for primary care providers to be able to recognize this disease so they can make prompt referrals to appropriate specialty centers.

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INTRODUCTION

lymphangioleiomyomatosis (LAM) is a relatively rare genetic disorder characterised by multiple lung cysts and progressive course that affects young women of childbearing age. It occurs as a sporadic disease (pulmonary LAM) or with tuberous sclerosis complex (TSC), which is inherited as an autosomal dominant disorder involving multiorgan hamartomas. [1] The disease is characterized by proliferation of unique smooth muscle-like cell (LAM cell) involving the small airways, pulmonary microvasculature, and lymphatic structures leading to airway obstruction, cystic changes in the lung parenchyma, pulmonary hemorrhages, chylous pleural effusions, and pneumothorax. [2] Caucasians are more commonly affected. [1] The prevalence of the disease affecting the lungs of premenopausal women is reported around one per million in the UK, France, and the US. [3] Reported prevalence in Asian countries is lower, around 0.24 and 0.03 per 100,000 of population in Singapore and Korea, respectively. [4]. LAM is exclusively confined to women, the mean age of onset being 34 years. [5],[6],[7] About 70% of LAM patients are 20-40 years of age at the time of onset of symptoms or diagnosis. Presentation after menopause is unusual. [7,9]

Case Report

A 68-year-old lady from maldives came to the emergency department with fever and progressive shortness of breath, orthopnea and cough with muco purulent expectoration for a period of 6 weeks. She Presented to maldivian hospital 6 weeks back with complaints of Fever, nasal discharge, sore throat, and dyspnoea and took treatment with medications including multiple antibiotics and supportive care during her stay in Maldives. She was referred here due to persisting dyspnoea. She gives no history of haemoptysis, Paroxysmal nocturnal dyspnea, joint pain or swelling, rash, swollen legs or any H/o contact with Tuberculosis in the past. She had complaints of progressive breathlessness for last 1 years. Initially it was exertional in nature and presently progressed to a state where patient is dyspneic at rest. Past History shows she is suffering from LAM with respiratory failure requiring intubation and ICU care 1 year back. She was recently detected to be a diabetic, 3 months back ted on oral anti diabetic agents. None of her family members had any h/o lung diseases, Tb, connective tissue disorder or malignancy. She did not complain of any loss of weight, alteration in bowel and bladder habits. She gives a history of smoking pipe at her 20, but stopped few years back. On physical examination, patient was well built and obese lady

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(BMI-34). All the vitals were in normal expect her Respiratory rate was 32/min and mildly dyspnoic. Patient was hypoxic and cyanosed, but improved with oxygen supplementation. She had Clubbing Grade 2 with Minimal soft pitting B/l Pedal edema +. She had no evidence of any neuro-cutaneous skin markers of tuberous sclerosis. Respiratory system examination was unremarkable except bilateral basal crepitations and diminished breath sounds. Cardiovascular system examination did not reveal any abnormality. Other systems were essentially normal. Her routine blood investigations showed leucocytosis with neutrophilia with elevated acute phase reactants. Antinuclear antibody (ANA) and human immunodeficiency virus (HIV) test were negative. Arterial blood gas analysis showed Chronic compensated respiratory Acidosis and O₂ saturation 68% Suggestive of type 2 respiratory failure. Chest sciagram [Figure 1] showed homogeneous opacity of both lower zone and mid zone (possible collapse consolidation and bilateral effusion) With Cardiomegaly and mediastinal enlargement, Echo Showed minimal Pericardial effusion otherwise unremarkable. She was started on empirical broad spectrum antibiotics, Non Invasive ventilation and supportive care. Sputum culture showed *Acenetobactor baumanii* sensitive to current antibiotic hence continued, NIV was tapered off slowly and shifted to room within 8 days.

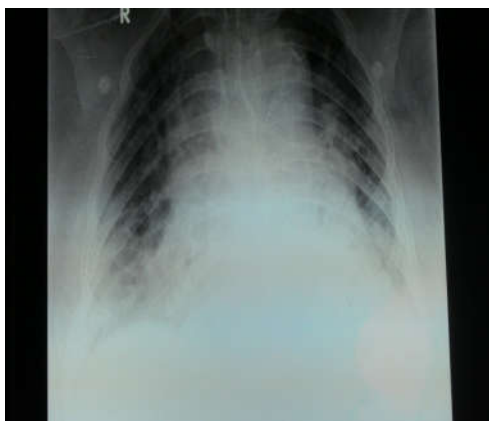


Fig 1 Chest sciagram showed homogeneous opacity of both lower zone and mid zone (possible collapse consolidation and bilateral effusion) With Cardiomegaly and mediastinal enlargement.

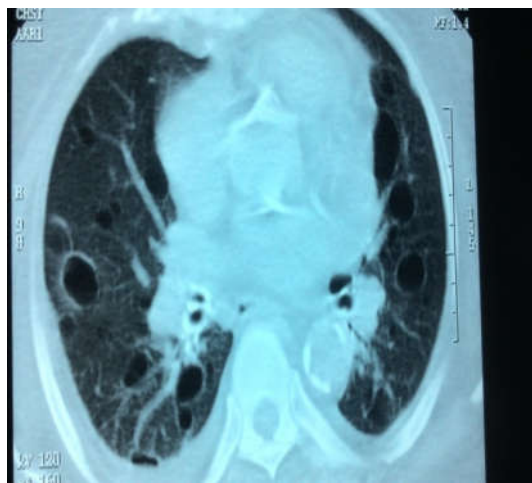
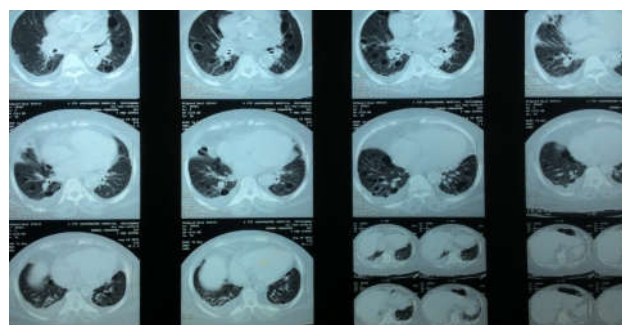


Fig 2 High resolution computed tomography (CT) thorax showed multiple thin walled cysts scattered bilaterally with areas of interseptal thickening, Sub segmental atelectasis of right lower lobe apical and basal segments with moderate pleural effusion in right side and mild on right side.

High resolution computed tomography (CT) thorax [Figure 2] showed multiple thin walled cysts scattered bilaterally with areas of interseptal thickening. It also showed Sub segmental atelectasis of right lower lobe apical and basal segments with moderate pleural effusion in right side and mild on right side. Spirometry showed obstructive pattern with increased lung volumes and reduced diffusion capacity (DLCO). In view of severe hypoxia, non-invasive procedures like bronchoscopy. The diagnosis was confirmed on the basis of classical high resolution CT findings. She was diagnosed with sporadic Lymphangioliomyomatosis with *Acenetobactor baumanii* sepsis with Type2 DM and Dyslipidemia. She showed good improvement in oxygen saturation and was discharged with oral steroids, antidiabetics, statins and domiciliary oxygen supplementation. The case was reviewed after 4months. There was significant improvement in dyspnea, spirometry, and arterial blood gas values. However, radiological features remained stationary with no deterioration.



DISCUSSION

A lung cyst is an air-filled lucent area with sharply demarcated thin walls (<3 mm). Cystic lung disease is characterized by multiple intrapulmonary cysts. LAM, a type of cystic lung disease is a relatively rare pulmonary disease of still unknown aetiology and pathogenesis. It is known that the disease progresses slowly until death in 8 to 10 years after diagnosis. It Usually manifests in the fertile stages of a woman's life, with exacerbations during menstruation and pregnancy, which indicates a probable hormonal influence. Diverse studies indicate that the progress of LAM may be accelerated by oestrogen and slowed by progesterone. But there are case reports of LAM occurring in post menopausal age group [8]. The clinical manifestations of LAM are dyspnea (59%), pneumothorax (49%), and cough (39%). Chest pain (22%), chyloous pleural effusions (13%), hemoptysis (18%), and wheeze are less common. [5],[6] Common extrapulmonary manifestations are retroperitoneal and renal angiomyolipomas. [9] Spontaneous pneumothorax can occur in 40-80% of patients and up to 50% experience kidney angiomyolipoma. Chylothorax or chyloous ascites can be associated with or without lung lesions.

Diagnosis of LAM is done by open lung biopsy (gold standard) or thoracoscopic biopsy based on characteristic histological findings. Immunostaining with HMB-45, specific markers for smooth muscle components namely actin and desmin increases the specificity and sensitivity. In advanced cases of LAM, open lung biopsy or transbronchial lung biopsies are avoided due to severe cardiopulmonary complications. But HRCT can often confirm the diagnosis and tissue diagnosis may not be

necessary. HRCT findings suggestive of LAM are small, thin walled, air containing cyst ranging from 2-20 mm or more in diameter scattered throughout the lung fields. There is no specific treatment or cure for this condition. However, oophorectomy, progesterone, tomosifen have been employed in the management of LAM.

Our patient presented with progressive dyspnoea which was diagnosed as chronic obstructive pulmonary disease and treated until 1 year back. She was diagnosed as LAM when she was admitted with respiratory failure one year back. She later failed follow up until presented now with progressive dyspnoea and Acinetobacter sepsis. She was started on tamoxifen, bronchodilators and domiciliary oxygen therapy prior to discharge. The diagnosis of LAM was made on the basis of the radiological findings.

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