

Spontaneous Pneumopericardium in a Patient of Lymphangioliomyomatosis and Pulmonary Tuberculosis- Rare Case Report

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Abstract

Pneumopericardium is a rare entity and is usually reported as a complication of trauma [1]. We are reporting a rare case of Lymphangioliomyomatosis (LAM) coexisting with sputum positive miliary tuberculosis presenting with pneumopericardium. LAM is a rare lung disease of unknown etiology, exclusively affecting women of reproductive age group and is characterised by proliferation and infiltration of pulmonary interstitium with atypical smooth muscle cell. LAM causes pleural complications especially pneumothorax and chylothorax, but it has not been reported to cause pneumopericardium. However pulmonary tuberculosis is known to cause pneumopericardium but coexistence of LAM with Pulmonary tuberculosis is a rare presentation.

Keywords: Pneumopericardium, Lymphangioliomyomatosis, Pulmonary Tuberculosis, Thoracoscopic drainage

Case Report

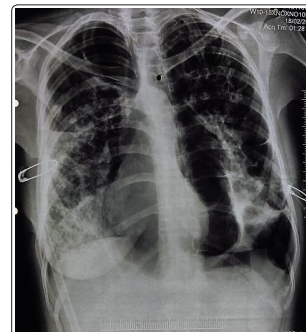
A female receptionist aged 25 years, non smoker, recently diagnosed to have miliary tuberculosis on anti tubercular therapy presented with complaints of fever and breathlessness for 3 days. On presentation in the Emergency department her Pulse was 120/minute, Blood pressure- 110/70 mmHg, Respiratory rate-24/minute and SpO₂ was 80% on room air. On auscultation breath sounds were absent in right infrascapular and inframammary area, elsewhere crackles were present. Arterial blood gas (ABG) analysis showed PH -7.48, P_aCO₂- 50 mmHg, P_aO₂- 58mmHg, HCO₃⁻-30mEq/L, at Fio₂ of 28%. Chest x-ray showed right pneumothorax with bilateral cystic changes and nodules. A right intercostal tube (ICD) was inserted following which the lung expanded and pneumothorax resolved and ICD was removed.

After one week, she developed pneumothorax on the same side, right ICD was reinserted. Simultaneously she developed left pneumothorax and subsequently left sided ICD was inserted. Contrast enhanced computed topography (CECT) chest done was suggestive of bilateral loculated hydropneumothorax with septae and partial collapse and destruction of underlying lung. The visualised lung showed irregular cystic changes and nodules (left>right), suggestive of cystic lung disease.

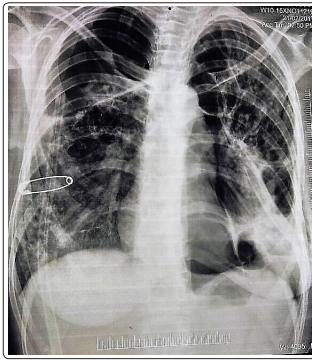
Fibreoptic bronchoscopy was done and bronchial washings were sent for Gene Xpert, which was positive for Mycobacterium Tuberculosis with no rifampicin resistance. Transbronchial lung biopsy, histopathology specimen features showed cystic dilatation of alveolar tissue with thickened septae due to the presence of spindle shaped smooth muscle cells and focal necrosis which was consistent with LAM. No active granuloma was seen. HMB 45 was negative indicating proliferation of spindle cell variety of LAM cell. Patient

was started on Sirolimus 1 mg per day with category 2 under DOTS (due to category 1 failure).

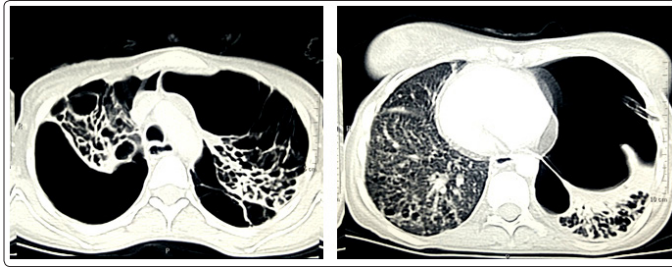
Two weeks later, patient developed sudden breathlessness and hypokalemia and chest x-ray done revealed a large pneumopericardium. As pneumopericardium was large causing mediastinal shift and compromising haemodynamic stability, CECT chest could not be performed. Percutaneous aspiration under echocardiographic guidance was attempted twice, but due to extensive cystic spaces within the lung and around the pericardium, procedure was unsuccessful. In view of poor respiratory reserve and bilateral cystic changes in the lungs, patient was considered to be unfit for general anaesthesia (GA). Hence it was decided to take up the patient for thoracoscopic guided drainage of pneumopericardium under local anaesthesia. Thoracoscope was introduced through the left intercostals tube drainage site. Pericardium was identified and a small incision was made over pericardium to drain the pneumopericardium followed by fenestration of the pericardium to prevent the recurrence. An ICD was placed from the same site as that of previous tube. Following the procedure patient improved symptomatically; pneumopericardium resolved which was confirmed by postoperative chest x-ray and subsequently the ICD was removed after 7 days.



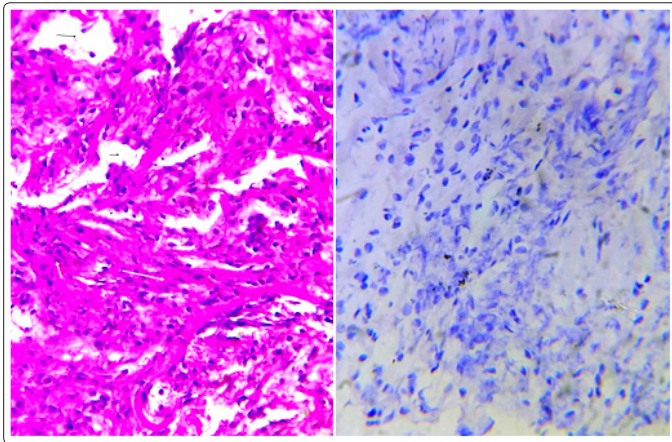
X- ray pneumopericardium



Post operative VATS



CECT Chest



Histopathology slides HMB 45 stain

Bricheteau first described pneumopericardium in 1844 with description of atypical “bruit de Moulin murmur”. Pneumopericardium usually occurs as a result of blunt trauma [2,3]. Various other causes are thoracocentesis, pericardiocentesis, thoracotomy, tracheostomy, endotracheal intubation, mechanical positive pressure ventilation, barotrauma, Valsalva and Heimlich manoeuvre, bone marrow sternalpuncture, endomyocardial biopsy, infected fluid or gas producing organisms in the pericardial sac, fistulous communication between pericardium and other air containing structures such as oesophagus, bronchus, stomach, amoebic liver abscess and Boerhaave syndrome. Other causes also includes foreign body aspiration, physical exertion, parturition, severe cough, acute asthma, cocaine inhalation, chlorine gas exposure, and forceful emesis [3-6].

Haemodynamic changes secondary to pneumopericardium mainly result from the volume and rate of air introduced into the pericardial cavity. Hemodynamic changes may occur with minimum of 60 ml of air introduced rapidly where as even 500 ml of air introduced

into pericardial cavity gradually, may be or may not be associated with major haemodynamic effects [7]. Since the patient developed sudden onset dyspnoea with hemodynamic instability the condition was considered to be rapidly progressive.

The possible mechanism of pneumopericardium in this case could be a severe bout of cough causing disruption of cystic lung spaces and the air tracking into the pericardium through a fistulous communication of pericardium with the diseased cystic lung [8]. Macklin in 1939 described the pathogenesis of pneumopericardium. According to him the rise in intra-alveolar pressure above atmospheric pressure resulting from excessive bout of coughing, lead to rupture of alveoli and the released air moves into hilar area, to the mediastinum, and through the pericardial reflections on the major pulmonary vessels into the pericardial cavity. The weakest histological area is parietal pericardium reflecting on to the visceral pericardium near ostia of the pulmonary veins [9]. It is likely that in this patient necrotizing pulmonary process (Tuberculosis) coexistent with LAM resulted in a direct fistulous communication between the lung and pericardium leading to pneumopericardium.

Many of the patients presenting with small pneumopericardium are asymptomatic and in them the pneumopericardium may resolve spontaneously. These patients are usually diagnosed incidentally on a chest radiograph and the level of gas in erect position is usually below the upper limit of the pericardium in the erect position, which differentiates it from pneumomediastinum [3]. However patients with large pneumopericardium are symptomatic and usually present with dyspnoea and precordial chest pain. Other symptoms are usually associated with the underlying etiology, as in this patient.

Pneumopericardium is usually self-limiting and does not require any specific treatment. However treatment is required in cases of large and symptomatic pneumopericardium or in patients with tension pneumopericardium. The treatment can be ECHO guided needle aspiration, tube decompression or pericardiectomy. In our case ECHO-guided multiple percutaneous aspirations failed to resolve the pneumopericardium. As she was considered unfit for general anaesthesia, she underwent thoracoscopic pericardiectomy under local anaesthesia. Fenestration was done to prevent recurrence. Oxygen therapy in high concentration was given in conjunction with surgical intervention to prevent retention of air. Chest x-ray at immediate post-operative period did not show any recurrence of pneumopericardium.

Conclusion

This case emphasises the utility of non intubated thoracoscopic drainage of pneumopericardium under local anaesthesia, when patient is considered unfit or at a very high risk for general anaesthesia.

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