



Bilateral spontaneous pneumothorax as the presenting feature in lymphangioleiomyomatosis

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Introduction

Bilateral pneumothorax is rare (1) and most commonly follows trauma or iatrogenic injury (2,3). While spontaneous pneumothorax is not uncommon, it is bilateral in less than 5% of cases (4). We report a case with bilateral spontaneous pneumothorax as the presenting symptom of lymphangioleiomyomatosis (LAM).

Case Report

A 46-year-old woman presented to the Hadassah University Hospital with progressive dyspnoea of 3 days duration. She had no accompanying fever, cough or chest pain. She had no previous illnesses or respiratory symptoms and had never smoked. On examination, respiratory rate was 25 breaths min^{-1} , pulse 88 beats min^{-1} and blood pressure 100/60 mmHg. There was no cyanosis. There was increased resonance to percussion and reduced breath sounds over both lung fields. Chest X-ray showed bilateral pneumothoraces, 40% on the left and 30% on the right. Bilateral intercostal drains were inserted with complete re-expansion of both lungs. No evidence of pulmonary parenchymal abnormalities was noted. Despite complete re-expansion, significant air leak persisted bilaterally and on the sixth day, chemical pleurodesis using 1.25 g of tetracycline hydrochloride in each hemithorax was performed. On the right side, the air leak resolved and the intercostal drain was removed. The leak on the left side, however, persisted.

On the twelfth day of admission, the right pneumothorax recurred and an intercostal drain was

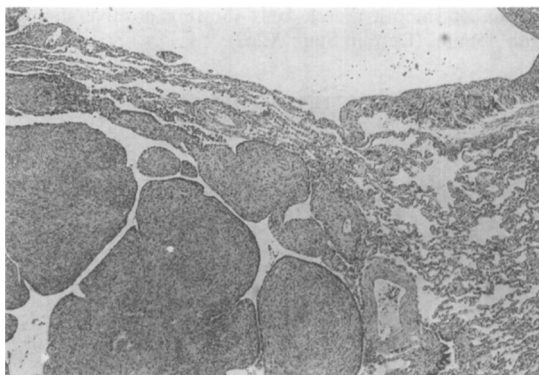


Plate 1 Low power photomicrograph showing nodules of smooth muscle protruding, and partially occluding the lumen of a bronchiole. Note the adjacent cystically dilated air space and disorderly proliferation of smooth muscle along the alveolar septa. (Hematoxylin – Eosin X26).

re-inserted. On day 20, because of persistent leaks on both sides, surgery was performed.

A midline sternotomy was performed to allow access to both hemithoraces. The lung surface showed multiple bullae in both lungs. The larger surface bullae were resected and then over-sewn. Wedge biopsies were taken and pleural abrasion was performed before surgical closure. There was no post-operative air leak and intercostal drains were removed 2 days after surgery. Pathological examination of the lung biopsies showed classic changes of LAM (Plates 1 and 2).

Pulmonary function tests showed normal volumes and flow rates with only a mild reduction in diffusion capacity (67% of predicted). Computerized tomography of the chest (Plate 3) showed multiple cystic changes in both lungs.

As the patient had no symptoms and only minimal findings on lung function tests, we decided not to administer further treatment. She has been followed for 36 months and has had no recurrence of

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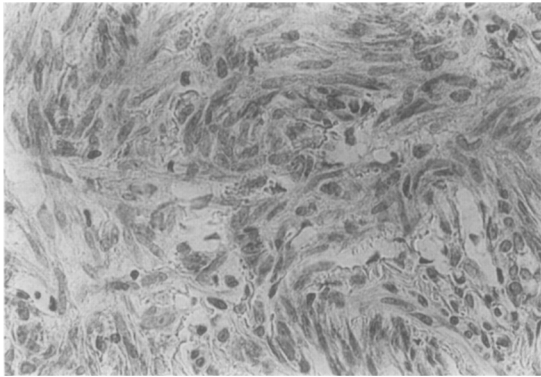


Plate 2 The nodules are composed of a disorderly proliferation of smooth muscle cells showing positive staining with desmin. (Desmin Stain X262).

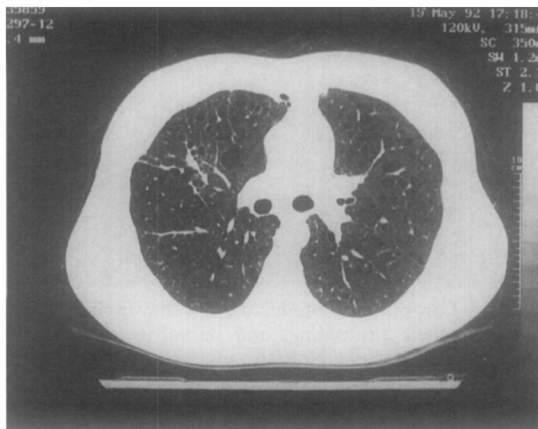


Plate 3 Thin section CT of the chest showing multiple, thin-walled cysts.

pneumothorax, nor has she developed other pulmonary symptoms. Follow-up CT 2 yrs after presentation showed a slight increase in cyst size.

Discussion

Spontaneous pneumothorax may be primary or secondary to underlying lung disease. Of 318 pneumothoraces reported by Melton *et al.* (4), 75 were secondary to trauma, 102 were iatrogenic and 141 were spontaneous. Of the latter group, 77 pneumothoraces were primary and 64 were secondary. Primary and secondary pneumothoraces were 6.2 and 3.2 times more frequent, respectively, in males than in females.

Simultaneous bilateral pneumothorax is uncommon, and most frequently follows trauma or iatrogenic injury (5). It may occur following venipuncture or catheterization of the internal jugular or

subclavian vessels (2), and has also been reported following tracheostomy and mediastinoscopy (3,5).

Spontaneous bilateral pneumothorax is rare, and accounts for less than 5% of spontaneous pneumothorax cases. Primary pneumothorax is seldom bilateral, with only four reported cases since 1983 (1). Bilateral spontaneous pneumothorax may also occur secondary to underlying lung disease, and has been reported in association with chronic obstructive lung disease, sarcoidosis, Marfans' syndrome and cavitating pulmonary metastases (1,6,7). Catamenial pneumothorax and pneumothorax in pregnancy have been reported to occur bilaterally.

LAM is a rare disease occurring exclusively in women, and by 1975, only 57 cases had been reported in the medical literature (8). A series of 32 cases, together with a review of the clinical and laboratory features of LAM and response to therapy, has recently been published (9).

Pathologically, LAM is characterized by smooth muscle proliferation within the walls of the pulmonary lymphatics, blood vessels and bronchioles. This results in lymphatic obstruction and chylothorax, vascular disruption with haemoptysis and airway obstruction with formation of multiple bullae and hyperinflation. The clinical picture is of progressive dyspnoea, pneumothorax, chylothorax and haemoptysis. In one series, pneumothorax was the presenting feature of LAM in 21% of cases, and developed at some time during the illness in 43% of cases (8). Others have found pneumothorax even more commonly: present in 53% of cases at presentation, and 81% during the course of the illness (9). It often occurs in the absence of other clinical or radiological features of the disease, is often recurrent and may be refractory to conservative tube drainage.

A significant number of patients with LAM develop pneumothorax in both chest cavities at different times during their illness.

While unilateral pneumothorax occurring spontaneously in a young, apparently healthy female with an otherwise normal chest X-ray is most likely to be idiopathic, the same does not apply for pneumothorax occurring bilaterally, and in these circumstances, LAM should be considered high amongst the diagnostic possibilities.

Once LAM is suspected, a diagnosis can be attempted by a transbronchial biopsy (9). If transbronchial biopsy is not diagnostic, or conservative management of the pneumothoraces is unsuccessful, open thoracotomy with open lung biopsy and pleurodesis should be performed. Early diagnosis of LAM allows close follow-up and institution of appropriate therapy if and when this becomes

necessary. The role of hormonal therapy is not completely clear and should probably only be given to patients who are symptomatic.

In conclusion, our patient presented with bilateral spontaneous pneumothoraces as the initial manifestation of LAM. Any unusual presentation of a young female with pneumothorax should raise the clinical suspicion of pulmonary LAM, despite normal chest X-ray and normal pulmonary function tests.

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