Recurrence Pneumothorax
In Young Women

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Introduction
This case is of interest as it highlights the presentation of a rare but serious condition which may be overlooked if knowledge of the presentation is not known.
The aim of this poster is to increase awareness of the condition so that it can be recognised and managed promptly and appropriately

Case
A 29 year old lady was admitted to Acute Medicine at QEHB from the emergency department with a left sided pneumothorax. She described sudden onset left sided chest pain and dyspnoea when she was in nursing.
Of note four days previously she had presented with right sided chest pain and dyspnoea. A chest X-Ray revealed a right sided pneumothorax which was aspirated with good effect. She was discharged home from the emergency department.
She had no significant past medical history, did not take regular medications and was fit and well. She was an office worker and had never smoked.
A more detailed history noted that she had been treated for a pneumothorax one year previously with aspiration.
A chest radiograph revealed a resolving right pneumothorax and a new left sided pneumothorax in otherwise normal looking lungs. (Figure 1)
The case was discussed in the daily handover meeting. The clinical lead for the unit recommended computerised tomography scanning to assess for any underlying abnormality.
The scan revealed multiple well-defined small cysts throughout both lungs, consistent with diagnosis of lymphangioleiomyomatosis. (Figure 2)
The patient was discharged home with information about the condition and with respiratory follow up.

Lymphangioleiomyomatosis (LAM)
Lymphangioleiomyomatosis is a rare, slowly progressive cystic lung disease affecting mainly women of child bearing age 1.
It is characterised by the proliferation of atypical pulmonary interstitial smooth muscle and epithelioid cells surrounding bronchovascular structures 2. (Figure 3) The mechanism of cyst formation is unknown.
Over time the normal lung parenchyma is remodelled and the cystic structure predisposes to pneumothorax 3. (Figure 4)
The pathogenesis of LAM is unknown, but data exists to suggest the loss of tumour suppressor function in specific cellular proteins. It is likely that oestrogen plays a role in disease progression given it does not occur prior to menarche, is exacerbated by pregnancy and symptoms improve following oophorectomy 4.
LAM usually presents with progressive dyspnoea and spontaneous pneumothorax. On examination end-expiratory crackles may be present, but examination is usually unremarkable. Investigations to aid diagnosis can be seen in the table below.

<table>
<thead>
<tr>
<th>Pulmonary Function</th>
<th>Mixed or obstructive pattern</th>
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<tbody>
<tr>
<td>Chest Radiograph</td>
<td>Initially normal but lung may become hyper inflated</td>
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<tr>
<td>Computerised Tomography</td>
<td>Diffuse, homogenous, thin-walled cysts scattered throughout both lung fields</td>
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<tr>
<td>Tissue biopsy</td>
<td>Stains specific for smooth muscle components</td>
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LAM is a progressive disease and will eventually lead to respiratory failure. The estimated median survival time in the United States is 23 years from diagnosis 5.

Treatment options are limited, there are no pharmacological treatments with proven benefit.
For some patients bronchodilator therapy may give symptomatic relief, but this does not alter disease progression. The use of hormonal manipulation and sirolimus has been trialled, but more information about their benefits is needed.
Lung transplantation offers an improvement in lung function and patient survival after lung transplantation is similar to that for pulmonary fibrosis and emphysema 6.

Key Messages
• A full and detailed history is the cornerstone of any clerking
• A handover meeting of all cases presenting to acute medicine promotes good practice within an acute medicine unit
• The majority of spontaneous pneumothoracies in young women are benign
• The British Thoracic Society guidelines do not provide guidance for investigation of recurrent pneumothorax
• Computerised tomography should be considered in all recurrent pneumothoraces
• Respiratory in-reach is an invaluable resource in an acute medicine unit
• Consider lymphangioleiomyomatosis as a diagnosis in recurrent pneumothorax

References