

Images In Medicine: Infected Giant Bulla, An Unusual Presentation Of Lynphangiomyomatosis

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Citation

C Padalino, C Sabella, F Toso, E Lugatti, C Paolillo, R Sbrojavacca. *Images In Medicine: Infected Giant Bulla, An Unusual Presentation Of Lynphangiomyomatosis*. The Internet Journal of Pulmonary Medicine. 2008 Volume 11 Number 1.

Abstract

Lynphangiomyomatosis (LAM) is a rare disease characterized by progressive cystic destruction of lungs. Giant bulla is rarely found in patient with LAM. We report an unusual presentation of LAM with infected giant bulla.

A 40-year-old female non-smoker presented to the ED with non productive cough and fever of 4 days duration. There was no associated chest pain, hemoptysis or dyspnea.

She had no significant medical history.

Her vital signs were as follows: blood pressure 110/60 mmHg, pulse 100 beats/min, respiratory rate 22 breaths/min and temperature 37,5°C. Oxygen saturation was 92% in room air.

There were decreased breath sounds particularly on the right side. The rest of the examination was unremarkable.

Laboratory analysis yielded leucocytosis (13.500 cells/ul) and increased PCR (250 mg/dl). Arterial blood analysis revealed that her PaO₂ was 55 mmHg and pCO₂ 32 mmHg.

Chest radiography showed bilateral hyperinflation and a large hypertransparent image with two fluid levels in the upper lobe of the right hemithorax.

A HRCT revealed multiple round thin walled cysts uniformly distributed throughout the lungs with minimal normal parenchyma and fluid containing giant cyst (size 15 x 10 cm), occupying the superior half of the right hemithorax. Pneumothorax and pleural effusions were not observed (Fig.1).

A lung biopsy was performed and the pathologic analysis enabled us to confirm the diagnosis of lynphangiomyomatosis (LAM).

{image:1}

LAM is a rare multisystem disorder that occurs predominantly in women of reproductive age and typically presents with progressive dyspnea or pneumothorax.^{1,2}

The peculiar presentation of our patients was a fever with radiologic evidence of infection in a large bulla within advanced diffuse cystic lung disease.

To our knowledge, infected giant bulla, common complication of pulmonary emphysema³, has not previously been reported in association of LAM particularly at clinical presentation.

References

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