Rare disease

Spontaneous pneumomediastinum and subcutaneous emphysema in systemic lupus erythematosus

AlaEldin H Ahmed,^{1,2} Elrasheid A Awouda²¹ Department of Medicine, University of Khartoum, Khartoum, Sudan² Elshaab Teaching Hospital, Khartoum, Sudan

Correspondence to AlaEldin H Ahmed, draahmed@hotmail.com

Summary

We report a case of a 29-year-old woman who is known to have systemic lupus erythematosus and idiopathic interstitial pneumonia; she presented with a 1-day history of substernal chest pain and increasing shortness of breath. On examination, she was found breathless, but was not distressed or afebrile or normotensive. Auscultation of the heart revealed a positive Hamman's sign. There was chest-wall crepitus, and auscultation of the lung showed bilateral crepitations. Full blood count and biochemical profile were unremarkable. Chest x-ray showed signs of idiopathic interstitial pneumonia in addition to pneumomediastinum (linear band of air within mediastinal planes and continuous diaphragm sign) and chest-wall subcutaneous emphysema. She was treated with high-concentration oxygen. A repeat chest x-ray 5 days later showed complete resolution of the pneumomediastinum and subcutaneous emphysema, but signs of idiopathic interstitial pneumonia continued to persist.

BACKGROUND

Spontaneous pneumomediastinum is an uncommon and usually self-limited condition.¹ It results from rupture of alveoli adjacent to the mediastinum and is caused by a variety of conditions, including straining against closed glottis, blunt chest trauma, asthma and interstitial lung diseases.² There are numerous radiological signs of pneumomediastinum, and these depend on delineation of the normal anatomical structures that are outlined by air within the mediastinal planes. Anatomically, the mediastinum communicates with several anatomical structures outside the chest, including the submandibular space,² the retropharyngeal space and the vascular sheath in the neck.² Also, mediastinal tissue planes extend to the retroperitoneal space and from there extend all the way to the pelvis.³ In the case of pneumomediastinum, air may escape into these planes, giving rise to further physical and radiological signs. We report a rare case where spontaneous pneumomediastinum with thoracic-wall subcutaneous emphysema occurred in a patient with systemic lupus erythematosus (SLE) and idiopathic interstitial pneumonia.

CASE PRESENTATION

A 29-year-old woman presented to hospital with substernal chest pain and increasing shortness of breath for 1 day. She is known to have SLE with idiopathic interstitial pneumonia diagnosed 8 months previously on the basis of the presence of pleuritis, arthritis, positive anti-nuclear antibody and anti-dsDNA. The patient was receiving oral prednisolone 20 mg/day. On clinical examination she looked ill, breathless, but was not in respiratory distress. Her oxygen saturation, breathing room air, was 95%. She was afebrile (temperature 36.6°C) but not tachycardic (pulse 92 beats per minute) or normotensive (blood pressure 110/70 mm Hg), and auscultation of the heart revealed

a positive Hamman's sign. Subcutaneous crepitus was detected on palpation of the chest wall. Auscultation of the lungs revealed bilateral crepitations. Rest of the physical examination was normal. Her full blood count and biochemical profile were unremarkable. The chest radiograph (figure 1) showed signs of interstitial lung disease caused by her known SLE; in addition it showed a linear band of mediastinal air (open arrows), continuous diaphragm sign (black arrows) and subcutaneous emphysema (white arrows). Spontaneous pneumomediastinum and subcutaneous emphysema were diagnosed. The patient received high-concentration oxygen, and a chest radiograph done 5 days later showed complete disappearance of the signs of pneumomediastinum and subcutaneous emphysema, but signs of idiopathic interstitial pneumonia continued to persist (figure 2).

OUTCOME AND FOLLOW-UP

Signs of pneumomediastinum and subcutaneous emphysema completely disappeared, but signs of idiopathic interstitial pneumonia continued to persist. The patient is under regular follow-up for her SLE.

DISCUSSION

Spontaneous pneumomediastinum has been described in patients with connective tissue disorders, along with interstitial lung diseases, including dermatomyositis and rheumatoid arthritis.^{4 5} Two cases of spontaneous pneumomediastinum and concomitant pneumothoraces have been described with SLE and idiopathic interstitial pneumonia.^{6 7} However, both patients died as a result of their illness. Our patient did not have a pneumothorax, and the pneumomediastinum resolved completely in 5 days. Another patient with spontaneous pneumomediastinum but without idiopathic interstitial pneumonia has been

Figure 1 Chest x-ray on presentation showing signs of interstitial lung disease, a linear band of mediastinal air (open arrows), continuous diaphragm sign (black arrows) and subcutaneous emphysema (white arrows).

Figure 2 Chest x-ray after treatment showing no signs of pneumomediastinum or subcutaneous emphysema but persistence of signs of idiopathic interstitial pneumonia.

described with SLE.⁸ These cases highlight a spectrum of presentations and varying prognosis of this condition.

In interstitial lung disease, the alveolar rupture that causes pneumomediastinum is thought to result from interstitial fibrosis, or pulmonary infarction caused by pulmonary vasculitis which is often a feature of connective tissue disorders.⁵ Respiratory tract infection has been described as a possible precipitating factor in some cases.¹⁻⁵ In our patient, however, there was no clear precipitating factor.

Spontaneous pneumomediastinum is usually self-limited, but a prolonged course going beyond 2 months and only responding to treatment with high-concentration oxygen has been described.⁵ Although no specific treatment for pneumomediastinum has been suggested in a published series,¹ we treated our patient with high-concentration oxygen to expedite recovery. High-concentration oxygen has been recommended as treatment for pneumothorax and has been helpful for subcutaneous and mediastinal emphysema. Pneumothorax, subcutaneous and mediastinal emphysema is reabsorbed into capillaries by diffusion along a partial pressure gradient caused by the sum of partial pressures exerted by water, carbon dioxide, nitrogen and oxygen. With breathing 100% oxygen, nitrogen is washed out of the blood, thus increasing the gradient for gas absorption and causing a four- to six-fold increase in the rate of gas absorption.⁹⁻¹⁰ We assume that the speedy recovery of pneumomediastinum and subcutaneous emphysema in our patient may have resulted from treatment with high-concentration oxygen; but, in the absence of a case-control study, we cannot be certain, as most cases recover spontaneously.¹

Learning points

- ▶ Spontaneous pneumomediastinum is a rare complication of idiopathic interstitial pneumonia associated with SLE that results from the rupture of alveoli adjacent to the mediastinum.
- ▶ The chest radiological signs depend on delineation of the normal anatomical structures that are outlined by air within the mediastinal planes.
- ▶ The extrathoracic physical and radiological findings in pneumomediastinum can be explained by air tracking along anatomical communications between mediastinal tissue planes and structures outside the chest.
- ▶ Spontaneous pneumomediastinum is usually self-limited, but treatment with high-concentration oxygen expedites air absorption from the mediastinum and subcutaneous tissues and speeds up recovery.

Competing interests None.

Patient consent Obtained.

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