Spontaneous pneumomediastinum (Hamman's syndrome): presenting as acute severe asthma

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Abstract

Spontaneous pneumomediastinum (SPM) is an uncommon finding and clinicians must consider this during their clinical evaluation. High degree of suspicion and appropriate investigations play key roles in early diagnosis and avoiding potential life-threatening complications. SPM usually presents without any comorbidities (primary) or due to an underlying pathology (secondary), such as underlying asthma, barotrauma, valsalva manoeuvre or an oesophageal rupture. Patients can have varying clinical symptoms; the majority will have subcutaneous emphysema and Hamman's sign (mediastinal crunching sound on auscultation). The prognosis of SPM is usually good with resolution in most cases, and it has a low recurrence rate. We report a case of SPM in a young 19-year-old male who presented with symptoms of acute severe asthma and who made a complete resolution with conservative management.

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Case presentation

A 19-year-old male university student with a background history of eczema and asthma presented with a day history of flu-like symptoms, acute breathlessness, unproductive cough, chest tightness radiating to his neck and vomiting. His asthma was well controlled and he had one previous exacerbation as a child requiring a brief overnight hospital admission. He was compliant with his inhalers and was a nonsmoker. On examination, he was tall, thin built, alert, oriented and talking in full sentences. Respiratory examination showed polyphonic wheeze and crepitations at the lower sternal area. Baseline peak expiratory flow rate was 550 l/min (predicted 600 l/min). Blood tests showed leucocytosis with predominant neutrophilia and normal electrolytes. ECG showed normal sinus rhythm and arterial blood gases on room air showed hypoxic respiratory failure (pH, 7.38; pCO_2, 4.5 kpa; pO_2, 6.5 kpa; base excess, 4.1 mmol/l; lactate, 2.7 mmol/l; and, HCO_3, 24 mmol/l). Chest X-ray (Figure 1) showed air in the mediastinum with normal lungs and no evidence of pneumothorax. He tested negative for flu.

He was treated conservatively with high-flow oxygen, nebulisers, antibiotics and antiemetics. Although his hypoxia and wheeze improved, he developed progressive neck swelling and clinical examination showed worsening surgical emphysema. He underwent a CT thorax with concurrent 1.5% oral contrast administration pre-scan (Figure 2). This showed no extra luminal contrast extravasation (no evidence of oesophageal perforation), but a large volume pneumomediastinum with air tracking through the neck fascial planes and subcutaneous emphysema in the left chest wall. There was further evidence of air in the posterior neck, thoracic soft tissues and epidural space.

He was treated with conservative measures and had 5 days of intravenous antibiotics for a presumed mediastinitis with complete recovery. A follow-up chest X-ray performed after 6 weeks showed complete resolution (Figure 3).

Discussion

Spontaneous pneumomediastinum (SPM) is a rare entity and is characterised by air leak into the mediastinum, not secondary to any underlying disease.[1] This process is self-limiting and was first described in 1819 by Laennec and in 1939 was further characterised by Hamman.[2] It is common in tall and thin males, and presents between the second to forth decade with an average age of 25 years.[3] There are a paucity of data regarding the annual incidence of this condition; however, in a retrospective study it has been reported to be approximately 1 in 30,000 attendances to the emergency department.[4] Secondary pneumomediastinum may occur following barotrauma, mechanical ventilation, illicit drug withdrawal symptoms, tracheobronchial injury, cardiothoracic interventions, frequent retching, vomiting, oesophageal rupture, acute respiratory distress syndrome, weight lifting, parturition and straining against a closed glottis (valsalva manoeuvres).[5–8] A total of 25% of patients presenting with SPM have a combination of asthma and episodes of coughing bouts.[5] The exact aetiology of this

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Figure 1 The pneumomediastinum (red arrows)

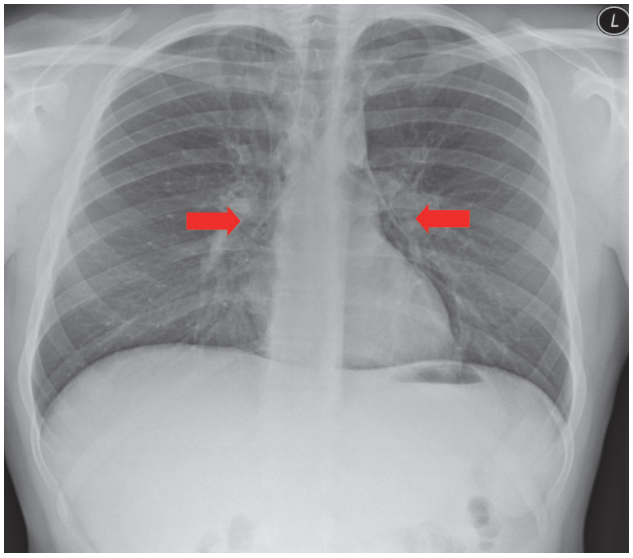
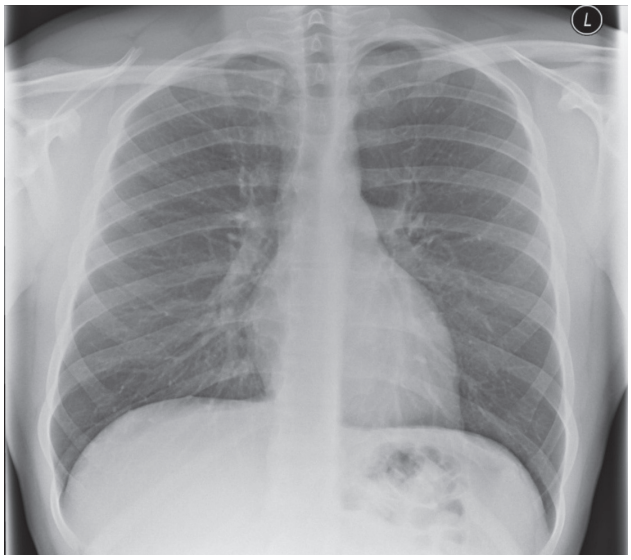


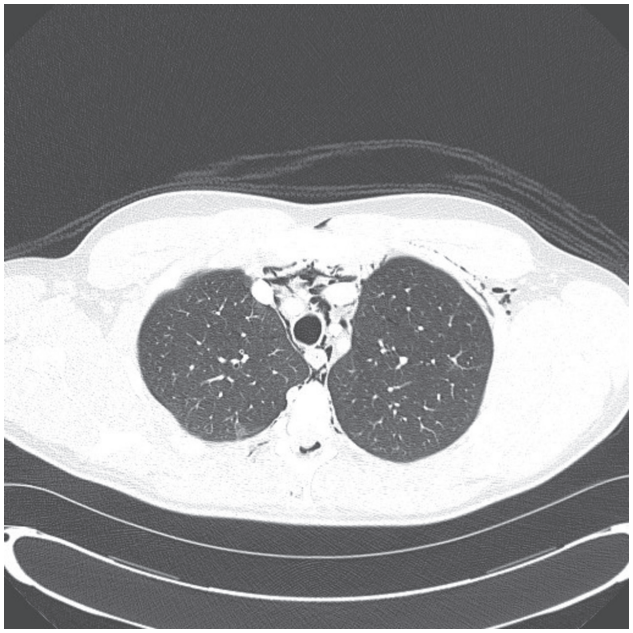
Figure 3 The resolution of the pneumomediastinum



process is unknown, but in patients with asthma SPM occurs secondary to air trapping due to airway narrowing or mucous plugging, especially after recurrent bouts of coughing. Intra-alveolar pressure increases leading to alveolar rupture causing the air leak to pulmonary interstitial spaces. There is extension of the air leak along the perivascular sheaths, dissecting into the mediastinum leading to mediastinal emphysema (Macklin effect).[5,9]

SPM presents with various symptoms, such as retrosternal chest pain (68%), neck pain and swelling, dyspnoea (44%), dysphagia and facial swelling.[10] Clinical signs include subcutaneous emphysema, and 'Hamman's sign' (mediastinal crunch that is characterised by precordial systolic crepitations synchronous with cardiac cycle) may be heard in about one-fifth of patients.[2,4,5] Our patient never had retrosternal chest pain but had evidence of 'Hamman's sign' on clinical examination. In a retrospective study, the majority of patients with SPM had subcutaneous emphysema (65%), Hamman's crunch (52%) and pneumothorax (11%).[3] Leucocytosis and elevated acute phase reactants, such as C-reactive protein, have been reported in 80% of patients with no evidence of localised or systemic infection.[10] Initial investigations should involve chest X-ray (anteroposterior and lateral views). SPM on chest radiograph is characterised by the presence of free air along the mediastinum or subcutaneous emphysema in the shoulders or neck region; however, chest radiographs underestimate the severity of SPM in up to 30% of cases.[11,12] CT scan with concurrent oral contrast is considered the gold standard diagnostic tool and is more sensitive. CT can detect oesophageal rupture and areas of air in the mediastinum or subcutaneous tissues that may be missed on a plain chest radiograph.

Figure 2 The pneumomediastinum and the extension of the subcutaneous emphysema in the neck and chest wall



SPM often follows a benign course and most patients diagnosed with uncomplicated SPM can be managed conservatively with advice to avoid any manoeuvres that increase the intrathoracic pressure. The possible underlying disease should be treated effectively to prevent recurrence or development of complications. The occasional complications encountered include pneumothorax (either unilateral or bilateral) requiring chest drainage, tension pneumomediastinum, pneumopericardium and cardiac tamponade.[4,5,13] Treatment with oxygen leads to rapid absorption of the air leak owing to an increase in the diffusion pressure of nitrogen.[5,13] Patients with SPM should be under observation to prevent further potential complications. SPM resolves with conservative measures with a good clinical outcome and has a very low recurrence rate provided the triggers are avoided, with one retrospective study reporting no recurrence rate over a 24-month follow up.[10] There are no recommendations or robust guidelines for follow-up radiological investigation unless patients have symptoms of recurrence or complications.

Conclusion

Although a rare phenomenon, clinicians should consider SPM during assessment of young adults with cardiorespiratory symptoms. Early diagnosis and high clinical suspicion prevents further complications and has a favourable outcome.

Informed consent

Written informed consent for the paper to be published (including images, case history and data) was obtained from the patient/guardian for publication of this paper, including accompanying images.

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