Recurrent idiopathic pneumomediastinum

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Summary: A case is described of the rare syndrome of recurrent idiopathic pneumomediastinum which had been mistakenly diagnosed as recurrent spontaneous perforation of the oesophagus. The case illustrates the importance of distinguishing between pneumomediastinum and oesophageal rupture because of their markedly differing prognosis and management.

Case history

In November 1978 a previously fit 19 year old warehouseman presented with chest pain and vomiting. He had been on a drinking spree 3 days earlier and had begun vomiting shortly afterwards. On the day of admission he had vomited blood on several occasions and he had developed chest and neck pain which was exacerbated by swallowing, retching and blowing his nose. On examination he was tall and thin with marked surgical emphysema of the neck extending down onto the chest. His body temperature was 37.4°C and heart rate 110 beats/min. Blood pressure was 140/90 mm Hg. A chest radiograph confirmed pneumomediastinum without pneumothorax or pleural effusion (Figure 1). A diagnosis of perforated oesophagus was suspected and he was immediately transferred to the Thoracic Surgical Centre. There a lipiodol swallow was found to be normal. He was managed conservatively by starvation, intravenous fluids and broad spectrum antibiotics. After 3 days when a barium swallow was normal, oral liquids were introduced uneventfully.

In August 1982 he was admitted to hospital with a history of repeated vomiting and haematemesis following an alcoholic binge. Chest radiograph showed pneumomediastinum. He was successfully managed conservatively.

In January 1984, he presented again with persistent vomiting of blood stained fluid. He complained of central chest pain exacerbated by swallowing and retching. He denied alcohol ingestion. On examination he was found to be pale and sweating with a temperature of 37.3°C. Heart rate was 90 beats/min and blood pressure 110/80 mm Hg. He had surgical emphysema palpable in the left supraclavicular fossa. Chest radiograph showed pneumomediastinum. His white cell count was 17.2 × 10⁹/l, and haemoglobin concentration was 16.2 g/l.

A diagnosis of perforated oesophagus was made and he was again transferred to the Thoracic Surgical Centre, where an immediate barium swallow showed mediastinal emphysema behind a normal oesophagus (Figure 2). He was managed as before and made an uneventful recovery.

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Figure 1 Chest radiograph showing mediastinal emphysema in tissue planes (arrows).
the pulmonary vasculature to the mediastinum and thence into the cervical and supraclavicular subcutaneous tissues (Rogers et al., 1972). The most common symptom is retrosternal chest pain which is exacerbated by coughing, swallowing, deep breathing and lying down but eased by sitting forward. Dyspnoea occurs in about half of the patients and usually abates with the appearance of surgical emphysema. About a quarter of patients are asymptomatic (Rogers et al., 1972). Clinically pneumomediastinum may be detected by loss of cardiac dullness and a characteristic crunching or crackling sound in time with the heart beat may be heard along the left sternal border described by Hamman (1939). Surgical emphysema which may take up to 12 hours to develop is usually felt in the neck, but can spread to involve the face, chest and abdomen.

The diagnosis is confirmed by chest radiograph which usually shows mediastinal emphysema at the level of the hila and cephalad within the mediastinum, and by the absence of oesophageal perforation on contrast oesophagography if performed within 24 hours of presentation.

Spontaneous pneumomediastinum is self-limiting and benign except when there is sufficient air in the mediastinum to compress the great vessels resulting in circulatory collapse (Macklin & Macklin, 1944).

Figure 2 Barium swallow showing mediastinal emphysema posterior to a normal oesophagus.

Subsequently a computed tomographic scan of the lung was normal with no evidence of bullae.

Discussion

A review of this patient’s history makes recurrent idiopathic spontaneous pneumomediastinum a more tenable diagnosis than oesophageal rupture. Spontaneous pneumomediastinum occurs in about 1 in 7,000 to 13,000 admissions in the USA (Yellin et al., 1983). It is associated with any condition in which high intrathoracic pressures are generated, i.e. vomiting, coughing, sneezing, asthmatic attacks, straining at stool or during childbirth or weight-lifting. It has occurred during general anaesthesia and during diabetic keto-acidosis. Women are as frequently affected as men with about half of the cases associated with parturition. It is thought to be due to rupture of alveoli at high pressure with dissection of the air along the course of the pulmonary vasculature to the mediastinum and thence into the cervical and supraclavicular subcutaneous tissues (Rogers et al., 1972). The most common symptom is retrosternal chest pain which is exacerbated by coughing, swallowing, deep breathing and lying down but eased by sitting forward. Dyspnoea occurs in about half of the patients and usually abates with the appearance of surgical emphysema. About a quarter of patients are asymptomatic (Rogers et al., 1972). Clinically pneumomediastinum may be detected by loss of cardiac dullness and a characteristic crunching or crackling sound in time with the heart beat may be heard along the left sternal border described by Hamman (1939). Surgical emphysema which may take up to 12 hours to develop is usually felt in the neck, but can spread to involve the face, chest and abdomen.

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Recurrent idiopathic spontaneous pneumomediastinum appears to be rare, being reported in one case after athletic activity (Yellin et al., 1983), in another during diabetic keto-acidosis (Nessan, 1974) and in a third in association with cyclical vomiting (Bullimore & Cooke, 1982). In each of these cases there was no serious outcome.

Because vomiting, chest pain and mediastinal emphysema are common to both conditions, spontaneous pneumomediastinum may be mistaken for spontaneous oesophageal rupture (Boerhaave's syndrome). However, the latter carries a poor prognosis with a mortality of 50% or more. Early diagnosis and surgical repair and drainage are advocated (Banks & Bancewicz, 1981). Mediastinal emphysema located posteriorly and inferiorly along the lateral edge of the aorta and beneath the parietal pleura over the left hemidiaphragm should suggest rupture of the oesophagus (Rogers et al., 1972).

Whilst patients with mediastinal emphysema clearly need immediate investigation and diagnosis it is important that the occurrence of spontaneous pneumomediastinum should be widely appreciated if unnecessary surgical or medical treatment is to be avoided.
References


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