Boerhaave's syndrome complicating status epilepticus

Introduction

Boerhaave's syndrome, first described in 1724, is a spontaneous transmural perforation of the oesophagus, typically caused by forced emesis. It is classically associated with the 'Mackler triad' of vomiting, lower thoracic pain and subcutaneous emphysema. It is the most lethal perforation of the gastrointestinal tract with an estimated mortality of 20–40% (de Schipper et al, 2009).

Discussion

Boerhaave's syndrome often presents a diagnostic challenge, the condition is rare and as in this case the classic triad of symptoms is not always present.

Other causes of subcutaneous emphysema include trauma or iatrogenic injury to the respiratory or gastrointestinal systems or from infection (gas gangrene). In a patient such as this one, who had recently undergone airway instrumentation and ventilation, potential causes of subcutaneous emphysema include hypopharyngeal or laryngeal perforation, or barotrauma secondary to high inflation pressures (Pan, 1989).

In this case subcutaneous emphysema was caused by air leaking through the perforated oesophagus into the mediastinum and decompressing along tissue planes resulting in subcutaneous air in the head and neck area, arms, chest and retroperitoneal space. The increased pressure in the mediastinum also appears to have ruptured the pleura and negative intrathoracic pressure drew air from the oesophagus into the pleural space.

A review of the literature suggests that early surgical therapy within 24 hours of diagnosis minimizes mortality (Kiev et al, 2007). However, patients who fulfill the Cameron criteria may be treated conservatively (Cameron et al, 1979). These criteria suggest that if the oesophageal rupture is well contained in the mediastinum, there is no pleural soiling and there is no clinical evidence of sepsis, then conservative management may be considered.

Conclusions

This case highlights the varied presentation of this potentially fatal condition and the importance of prompt diagnosis and appropriate early management to improve outcome. BJHM

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Cameron JL, Kieffer RF, Hendrix TR et al (1979) Selective nonoperative management of contained intrathoracic esophageal disruptions. *Ann Thorac Surg* **27:** 404–8

de Schipper JP, Pull ter Gunne AF, Oostvogel HJM, van Laarhoven CJHM (2009) Spontaneous rupture of the oesophagus: Boerhaave's syndrome in 2008. *Dig Surg* **26:** 1–6

Kiev J, Amendola M, Bouhaidar D, Sandhu BS, Zhao X, Maher J (2007) A management algorithm for esophageal perforation. *Am J Surg* **194:** 103–6

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Figure 1. Chest X-ray shows subcutaneous emphysema throughout with bibasal pneumothoraces.

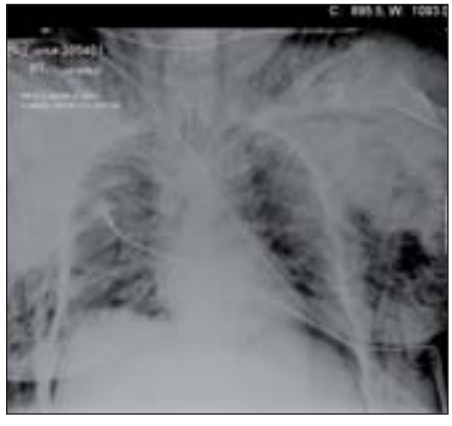
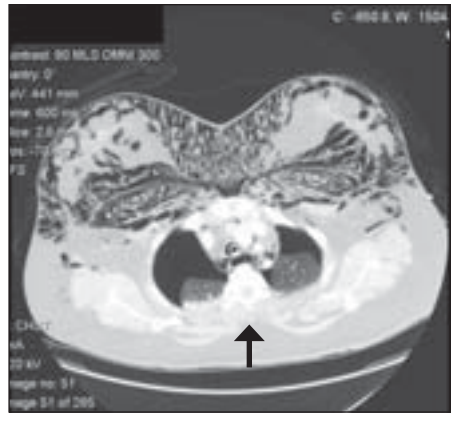


Figure 2. Cross-section of the thorax showing subcutaneous emphysema of the chest wall, bilateral pneumothoraces and pneumomediastinum. An oesophageal perforation is evident in the middle third of the oesophagus (arrow).



Case Report

A 40-year-old woman with a history of absence seizures presented to the accident and emergency department in status epilepticus following generalized tonic clonic seizures. The patient was drowsy (Glasgow Coma Scale 10/15) and had vomited several times during the preceding 24 hours. The seizures were controlled with lorazepam and phenytoin. She was sedated and intubated uneventfully and transferred to the radiology department for a computed tomography brain scan. The computed tomography scan of the brain was normal; she had no further seizures and was extubated, with a Glasgow Coma Scale of 12/15 post extubation. She had two further episodes of vomiting following extubation.

Two hours later she developed respiratory distress associated with a high-pitched voice, stridor, and massive facial and upper body swelling. She was intubated once again, this time there was a limited view of the airway secondary to soft tissue swelling. Chest X-ray and computed tomography of the thorax revealed massive subcutaneous emphysema and bilateral pneumothoraces (*Figures 1* and 2), which were decompressed. The computed tomography scan also revealed a focus of air within the mid-third of the oesophagus representing an oesophageal perforation (*Figure 2*). The patient was managed conservatively with intravenous antibiotics and nasojejunal feeding. Ten days later she was weaned from mechanical ventilation and transferred to the ward. She was discharged home 1 month after admission.