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A Rare Case of Reversible Bulbar Palsy Secondary to Boerhaave's Syndrome

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Introduction

Boerhaave's syndrome (BS) consists of a spontaneous longitudinal transmural rupture of the oesophagus, usually in its distal part. It generally develops during or after persistent vomiting as a consequence of a sudden increase in intraluminal pressure in the oesophagus. It is a rare but potentially life-threatening condition. The classic Mackler's triad of vomiting lower thoracic pain and subcutaneous emphysema is found in one third up to 50% of cases. Hematemesis is also an uncommon but yet challenging presentation. In comparison with ruptures of other parts of the digestive tract, spontaneous rupture is characterized by a higher mortality rate [1], dictating prompt diagnosis and treatment. We herewith report a case of BS with an unusual reversible neurological complication, the Collet-Sicard syndrome.

Case Report

A 30-year-old male patient was admitted to our hospital with acute onset of abdominal pain and vomiting for two days after he had eaten a large takeaway meal. His abdominal pain started in the epigastric area as initially colicky, and then moved centrally before becoming generalised. He had vomited more than twenty times with gastric contents before attending the A&E department, and presented with pyrexia and hyporexia. His past medical history included oesophageal strictures possibly related to eosinophilic oesophagitis. He was a moderate smoker (10-15 cigarettes per day) with occasional social alcohol intake. On admission, his observations were the following: BP 141/70 mmHg; HR 108 bpm; SO2: 95% on nasal cannula with 3L of oxygen. Blood Gas Analysis showed pH 7.380; pO2 7.79; pCO2 5.47; HCO3 24.3; Lactate 1.5. Admission Blood tests revealed Hb 175 mg/L WBC 15.0 10^9/L; Neutrophils 13.3 × 10^3/mL; PT 10.5, Na 140 mEq/L, K 4.7 mEq/L, Cr: 83, ALP 44 IU/L; Bilirubin 16 mg/dL; Amylase 140 U/L; CRP 6 mg/L; Troponin T 3 ng/ml.

Abdominal and Chest XR showed no gross abnormalities but the image quality was suboptimal. In contrast, a CT of chest, abdomen and pelvis showed extensive air in the mediastinum extending into upper abdomen with emphysema (Figure 1), which was associated with a developing bulbar palsy (Figure 2). CT with gastrografin showed extravasation of contrast in mediastinum, which was indicative of oesophageal perforation [2]. Patient was admitted to ITU and had a diagnostic laparoscopy with washout and placement of drains on the same day of admission. The following day he underwent an OGD with insertion of a fully covered oesopgaheal stent. Unfortunately, he required a prolonged stay in ITU due to sepsis caused by mediastinitis/pneumonia, type 2 Respiratory failures and difficult extubation and cuff leak with difficult reintubation and prolonged hypoxia for approximately 70 minutes. He also required a tracheostomy. Patient received enteral nutrition with NG tube until the oesophagus was fully healed. On discharge day, he had some residual dysphagia related to neuromuscular palatal and tongue dysmotility.

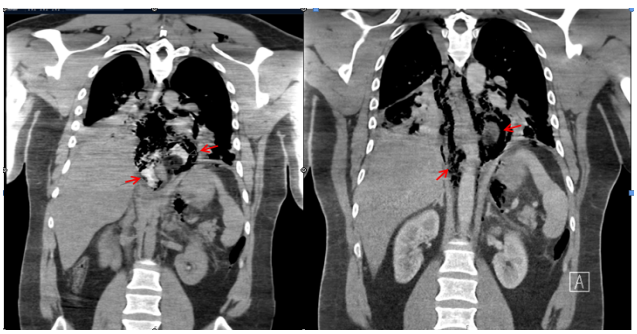

Figure 1: CT Chest/Abdomen: appearances suggestive of extravasated contrast in mediastinum.

On neurological review, he was diagnosed with Collet-Sicard syndrome [3] with resolving paralysis of the left sided 9th, 10th and 12th cranial nerves. A subsequent MRI of his head showed no abnormalities and an ENT review confirmed normal function and mobility of his vocal cords.

Results and Discussion

Spontaneous transmural rupture of the oesophagus was first reported in 1724 by Herman Boerhaave in Leiden [4]. Despite its benign etiology, the syndrome has a high mortality rate of 20-50% [5,6].

The pathophysiological mechanism involves a sudden rise in intraluminal oesophageal pressure following intense retching and vomiting, which results in rupture [1,2]. The majority of perforations occur in the distal oesophagus, typically on the left posterolateral wall,

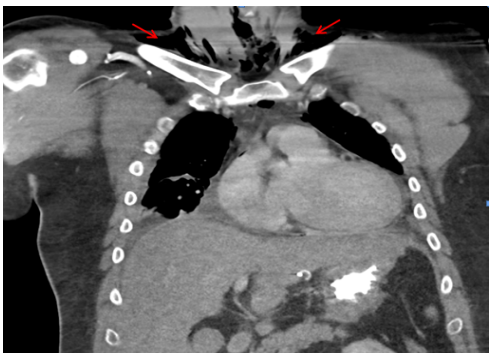

Figure 2: CT Chest: Neck emphysema that contributed to bulbar palsy.

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just above the diaphragmatic pinch, where an area of weakness has been demonstrated by anatomical studies [7,8].

Other concomitant factors favouring spontaneous rupture are the lack of serosa in oesophagus and in some patients, the lack or reduced presence of *Muscularis mucosae*.

It accounts for only 15–30% of all oesophageal perforations, with all other being secondary to endoscopic therapy, trauma, foreign bodies, or local oesophageal or pulmonary pathology [9,10].

Physical examination typically reveals subcutaneous emphysema and development of hydropneumothorax, usually on the left side.

The gold standard for diagnosis is an oesophagogram with a water-soluble contrast. However, CT is considered the most effective method for early detection of oesophageal perforation.

Diagnosis of BS can be difficult because history and symptoms are highly deceiving and nonspecific. Common symptoms include vomiting, chest pain, dyspnoea, dysphagia, subcutaneous emphysema, tachycardia, tachypnea, fever, and epigastric or abdominal pain. The differential diagnosis includes pulmonary embolism, myocardial infarction, aortic dissection, Mallory-Weiss tear, strangulated diaphragmatic hernia, pancreatitis and perforated peptic ulcer.

Surgical intervention is the mainstay of management, with the new advances in therapeutic endoscopy increasing interest in non-surgical options, including placement of a temporary fully covered oesophageal stent, Over the Scope Clip (OTSC), and endo-suturing.

Oesophageal resection/chest drain with or without oesophageal repair leads to survival up to 95% when treatment is provided timely within 24 hours.

Conservative management with NJ tube/TPN, antibiotics, and PPIs may result in 20-22% mortality.

Endoscopic management can be useful for perforations smaller than 10mm. The aim of endoscopic stenting is to prevent further extraluminal septic contamination and guide re-epithelialization of the oesophageal mucosa. Known complications of stent placement include stent migration, pressure ischaemia leading to ulceration/perforation, and reflux oesophagitis.

In a systematic review of 25 studies including 267 patients with complete follow up after temporary stent placement for benign rupture or anastomotic leak of the oesophagus, clinical success was achieved in 85% of patients and there was no difference between stent types (plastic stents 84% vs fully covered metal stents 85% and partially covered metal stents 86%, P=0.97). Time of stent placement was longest for plastic stents (8 weeks) followed by fully covered and partially covered stents (both 6 weeks). In 34% of cases there was stent-related complication. Stent migration occurred more often with plastic stents (31%) and fully covered stents (26%) than with partially covered stents 12%, whereas there was no significant difference in tissue in- and overgrowth between plastic and metal stents [11].

In a recent case report of BS from Portugal [12] a "damage control" stepwise approach was described, where a first surgery is performed to control sepsis with debridement and drainage of mediastinum and pleural cavity, followed by a second surgery, where the oesophagus is repaired.

The rationale for this approach is to prevent recourse to oesophageal resections with often high (up to 50%) postoperative leaks rates after primary repair [13]. The main disadvantage is that two thoracotomies and prolonged ICU stay are required. The main advantage is to better control the septic contamination whilst optimising the patient's conditions in preparation of the second operation.

In our case, we used a similar approach but instead of performing a second surgery, we chose an endoscopic treatment to seal up the perforation.

Despite optimal treatment, the patient developed an unusual complication, possibly related to hypoxia. The Collet-Sicard syndrome is a rare condition involving the base of the skull affecting both jugular foramen and hypoglossal canal resulting in unilateral and combined affection of lower cranial nerves (IX, X, XI, XII). It was first mentioned in 1915 during the World War One and related to bullet injuries [3,14].

Causes include tumour, vascular lesions, trauma, iatrogenic complications and inflammatory processes.

Patients present with a loss of taste in the posterior third of the tongue (IX nerve), paralysis of usually one (unilateral) vocal cord and dysphagia (X nerve), weakness in sternocleidomastoid and trapezius muscle (XI nerve), and atrophy and paralysis of tongue muscles (XII nerve).

With this case report, we would like to raise awareness of this rare neurological syndrome as a potential complication or consequence of Boerhaave's syndrome and its treatment.

Conclusion

Notwithstanding the delayed presentation, this case of Boerhaave's syndrome was promptly recognised and managed in a timely manner as much as possible. A minimally invasive approach with thoracic lavage and drainage was initially used to minimise septic contamination and the perforation was then sealed with a fully covered metal stent. However, a rare neurological syndrome developed possibly due to hypoxia, which reversed once the perforation was fully controlled.

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