

Boerhaave's Syndrome: 3 cases in 3 weeks in Remote Australia. A Case Series.

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Citation

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Abstract

Boerhaave's syndrome, a rare condition, not only poses a diagnostic and management dilemma¹, but also has a high morbidity and mortality. In remote Northern Territory, we saw three cases in three weeks in March 2009. These patients underwent surgery in our hospital. We present our series of three cases, our management and lessons we learnt.

INTRODUCTION

Boerhaave's syndrome, a rare condition, not only poses a diagnostic and management dilemma¹, but also has a high morbidity and mortality. In remote Northern Territory, we saw three cases in three weeks in March 2009. These patients underwent surgery in our hospital. We present our series of three cases, our management and lessons we learnt.

CASE SERIES

CASE 1

A 48-year-old indigenous gentleman presented to a remote clinic with vomiting, epigastric pain and sudden-onset left-sided chest pain after 3 days of binge drinking. He was a known heavy alcohol drinker but had no other medical comorbidities. He was febrile with a temperature of 37.8 degrees Celsius. He had decreased air entry to his left lung, had a respiratory rate of 50 breaths per minute and was saturating at 90% on room air. His abdomen was tender in the epigastrium with guarding. His heart rate was 120 beats per minute with a blood pressure of 120/80mmHg. Chest X-ray showed left pleural effusion. Haemoglobin, white cell count, urea and electrolytes and C-reactive protein (CRP) levels were all within normal limits. Liver function tests revealed a raised gamma-glutamyl transferase three times the normal levels. Lipase was 463 U/L (23-300).

He had an intercostal drain inserted on the left side and was evacuated to the Royal Darwin Hospital with a working diagnosis of pancreatitis with large pleural effusion 24 hours after presentation. On arrival at our hospital, he began to drain food particles in his chest drain whilst being assessed in the Emergency Department. A diagnosis of possible

Boerhaave's syndrome was made. He had a gastrograffin swallow and a computed tomography (CT) scan of his neck to assess the level of mediastinal contamination, which showed left pleural effusion, pneumomediastinum and an oedematous area in the lower oesophageal segment. He underwent a left thoracotomy (via sixth intercostal space), chest toileting and repair of a 4cm rent in the distal end of his oesophagus just above the diaphragm. He also had a feeding jejunostomy via a mini-laparotomy at the same time. Two chest drains were inserted; one anterior apical and the other posterior basal.

A Gastrograffin® swallow on post-operative day eight showed a small leak (see figure 1).

Figure 1

Figure 1. Arrow showing a small distal oesophageal leak on post-operative day 8.



This was managed conservatively. He made a slow recovery

requiring three weeks of ICU stay after which he developed a left-sided empyema. Three weeks post presentation he underwent a second thoracotomy and left lung decortication. He was subsequently discharged to the ward and he went home five weeks post presentation.

CASE 2

A 45-year-old Caucasian gentleman presented to Royal Darwin Hospital with severe epigastric pain radiating through to his back and three episodes of vomiting. He had been drinking heavily a day prior to presentation. He had a background history of active Crohn's disease for which he had had a right hemicolectomy two years prior to presentation. He was on prednisolone and azathioprine. On examination he had decreased air entry to his left chest with a respiratory rate of 22 breaths per minute. He was tender in his epigastrium. Chest x-ray showed small left pleural effusion. Abdominal x-ray showed a sentinel loop of dilated small bowel. He had a white cell count of $12.8 \times 10^9/L$ (4.0-11.0) and a lipase of 5600 U/L (23-300).

He was managed as severe pancreatitis. A CT scan of his chest showed a left pneumo-hydrothorax with pneumomediastinum. A Gastrografin® swallow showed a lower oesophageal perforation (see figure 2).

Figure 2

Figure 2. Gastrografin® swallow showing lower oesophageal perforation



He underwent a left thoracotomy (via 7th intercostal space). The finding was a 6cm rent abutting the hiatus. A

nasojejunal tube was inserted endoscopically via the tear with the aid of a paediatric gastroscope and a guide wire. The out-pouching mucosa was repaired and a Thal's patch gastroesophagoplasty was used to reinforce the repair. Two chest drains were inserted; anterior apical and posterior basal. Four days post-operatively, he developed a right-sided pleural effusion treated with a chest drain. Six days later, he developed a non-ST-elevation myocardial infarct in ICU. He was discharged home four weeks after presentation.

CASE 3

A 26-year-old Caucasian gentleman presented to the Royal Darwin Hospital with sudden onset of severe epigastric and chest pain after violent vomiting for three hours. He had been binge drinking. All his observations were normal. On examination, he had bilateral surgical emphysema of his anterior chest wall and neck. He was mildly tender in his epigastrium. All his blood results were normal. There was a high index of suspicion of oesophageal rupture. He underwent a CT scan of his chest which showed surgical emphysema and pneumomediastinum. A Gastrografin® swallow did not show any leakage of contrast.

He was managed conservatively (non-operatively) on intravenous antibiotics and nil by mouth for three days. Clear fluids were introduced and he was gradually started on normal diet. He was discharged after five days in hospital. His admission was uneventful.

DISCUSSION

Boerhaave's syndrome, a spontaneous oesophageal perforation, remains a difficult diagnosis due to its variable presentation². Recommendations regarding management are not clear cut and even sometimes controversial. The clinical manifestations of the disease are variable and often atypical⁴, and may be misleading, thus delaying early diagnosis. Some common presenting symptoms were reported by Brauer et al. in a series of 18 cases. These were; pain 83%, vomiting 79%, dyspnoea 39%, gastroduodenal ulcer 41%, and alcoholism and heavy drinking 40%³. Eroglu et al. reported fever in 33.3% of cases and subcutaneous emphysema in 65% of cases in their series of 36 patients⁵. We found vomiting and sudden-onset chest pain in the two patients we operated on. Diagnosis of oesophageal perforation can be suspected on chest x-ray. Pate et al. reported in their series of 24 patients over a thirty-year period, that 97% of chest x-rays were abnormal. Our two patients who required surgery had abnormal chest x-rays. An oesophagram was diagnostic in 23 out of 24 of their patients (95.8%)⁶. Dodd et al.

recommended a water-soluble contrast (Gastrografin®) as the medium of choice since barium induced inflammatory response in the pleural cavities, though it was superior in defining small perforations⁸. The test should be repeated with barium if Gastrografin® fails to define the perforation^{8,9}. All our patients had a Gastrografin® swallow and all those with contrast leak underwent surgery. CT scans play a vital role in diagnosis confirmation and may show suggestive features of oesophageal perforation when an oesophagram is normal⁷. All our patients had CT scans of the neck, chest and abdomen. This was important to assess the extent of contamination prior to surgery. The surgical management of Boerhaave's syndrome includes non-operative treatment, endoscopic (intraluminal) stent placement, operative surgery with primary repair with or without reinforcement, T-tube drainage, mediastinum drainage only, exclusion and diversion, and resection⁹. All these patients, irrespective of the method of treatment chosen, must have aggressive resuscitation and be admitted to an intensive care unit. In our case, two of our patients underwent left thoracotomies, one had a primary closure without reinforcement whilst the other had a Thal's patch gastroesophagoplasty. The third patient had intravenous antibiotics and close observation for five days.

CONCLUSION

A number of lessons were learnt from our experience. The symptoms of vomiting and sudden-onset chest pain, in addition to an abnormal chest x-ray, which we describe here as the "Darwin triad", were highly suggestive of oesophageal rupture. Extravasation of contrast into the thoracic cavity was essential in deciding whether our patients were managed conservatively or operatively. Seventh space thoracotomy gave us better exposure than sixth space thoracotomy which was further improved by

subperiosteal excision of the most posterior one centimetre of the rib. The use of a paediatric gastroscope was useful in the placement of a nasojejunal tube via the oesophageal tear using a guide wire. Small contrast leaks beyond three weeks of the primary operation in an otherwise stable patient with no pleural collection can be managed conservatively.

Rural surgeons with expertise should consider early surgical intervention in their hospitals if patient travel to regional hospitals is likely to be delayed.

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