

Short Report

Boerhaave's syndrome: Experience with patients presenting later than 24 hours

Amit Ganguly, Manish Porwal, Jagdish Khandeparkar

ABSTRACT

Department of Surgical
Gastroenterology and Laparoscopic
Surgery¹,
Cardio-Vascular Thoracic Surgery²
CHL–Hospital,
Indore, India

Boerhaave 's syndrome is the most sinister cause of esophageal perforation. Clinical presentation is vague. Diagnostic delays are frequent. As condition is rare therefore no consensus exists on management. A wide variety of management options are described in literature, each with its advantages and disadvantages. We present our experience of managing these cases which presented after 24 hr. Of onset of symptoms with emphasis on primary reinforced repair as first line surgical option.

Correspondence:

Dr Ganguly Amit

Email: dramitganguly@rediffmail.com

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Introduction

Boerhaave's syndrome (BS) or spontaneous esophageal perforation is an uncommon disease entity. Though it is called spontaneous, it is invariably due to barotrauma. The adjective spontaneous signifies that it not secondary to trauma, instrumentation or foreign body.¹ The usual mechanism believed responsible for perforation is sphincter muscle incoordination, which causes failure of the cricopharynx to relax, resulting in sudden increase in intra-luminal pressure in the esophagus. The perforation usually occurs in the left posterolateral aspect of the distal esophagus, which is considered the area of weakness.²

We present our experience of management in patients of BS presenting 24 hours after onset of symptoms. During the period of 2010 -2014, we treated 5 patients of spontaneous

esophageal perforation. The clinical features, contrast enhanced computerized tomography (CECT) findings, treatment and outcome are summarized in the **Table 1**.

Case report

Two patients (Patients 1 and 2) were resuscitated and taken up for surgery within six hours of admission. Both underwent left thoracotomy with primary reinforced repair. Post operatively they were electively intubated for 24 hours. A feeding jejunostomy was initiated after 48 hr. Gastro-graffin study was done on 8th post operative day. Patient 1 showed leak, which resolved without any intervention. His left intercostal drainage (ICD) was removed after resolution of the leak. All through this

Table 1: Patients with Boerhaave's syndrome: features and outcome

S. No.	Age/sex	Duration of illness	Clinical feature	CT scan findings	Management	Outcome
1.	42y/M	Two days	<ul style="list-style-type: none"> Chest pain, breathlessness & facial Puffing O/E- subcutaneous emphysema, Decreased air entry on Lt > Rt. 	B/L pleural effusion with air collection near distal esophagus with contrast extravasation in pleural cavity.	Primary reinforced repair with pleural patch with drainage of both pleural cavity & feeding jejunostomy.	Minor leak on Gastrograffin study, healed without any intervention.
2.	77y/M	Three days	<ul style="list-style-type: none"> Chest pain, fever and breathlessness O/E- Decreased air entry on Lt. Side 	Lt. Pleural effusion with collapse of Lt. lung with contrast extravasation in Lt. pleural cavity	Primary reinforced repair with pericardial fat pad with drainage of Lt pleural cavity & feeding jejunostomy	Uneventful.
3.	73y/M	Six days	<ul style="list-style-type: none"> Chest pain , fever O/E-decreased air entry on Rt. Side. 	Rt.pleural effusion, Rt paracardiac collection with contrast from esophagus extending in to collection.	Conservative* (Intercostal tube Drainage with feeding jejunostomy)	He was discharged and readmitted with hepato-renal syndrome. Died of Multi-organ failure.
4.	80y/F	Five days	<ul style="list-style-type: none"> Chest pain, breathlessness O/E- decreased air entry on RT. Side 	Rt. Pleural effusion and contrast extravasation	Inter costal Drainage, adv- stenting	Left against medical advice
5.	43y/M	One day	<ul style="list-style-type: none"> Pain following vomiting o/ E - NAD 	Wall thickening, intramural perforation and periesophageal collection.**	stenting	Uneventful.

* offered both stenting and conservative management .

** Under went Upper G.I. Endoscopy Before CT

period he was on feeding jejunostomy feeds. The mean duration of hospital stay for this group of patient was 2 weeks.

Patient 3 was HbsAg +ve with deranged INR, and low platelet count. Imaging revealed small shrunken liver, moderate ascites and collateral at porta. Surgery was deemed high risk. Intercostal drainage tube insertion and feeding jejunostomy were performed under local anesthesia. He was also offered endoscopic stenting but the family refused. He was discharged with intercostal drainage tube and feeding Jejunostomy in-situ. He had to be readmitted after 6 weeks with hepatic decompensation and azotemia. He died of multi-organ failure.

Patient 4 was an elderly lady, who at the time of diagnosis was not fit for any operative intervention. Intercostal drainage tube was inserted at the bed side. She was advised stenting but declined and left against medical advice.

Patient 5 was first seen in the out patients' department before being referred to us, where he presented with complaints of pain in epigastrium after a bout of vomiting. Acid peptic disease was suspected. At the commencement of upper gastrointestinal endoscopy (UGIE), suspicious perforation in the distal esophagus was identified. Endoscopy was terminated

immediately. The patient was admitted and CT scan done. In view of the minimal peri-esophageal mediastinal collection he was offered stenting. He was discharged after 5 days and subsequent CT scan at 6 weeks showed resolution of collection.

It is our policy to offer surgery to all patients if contrast-enhanced CT scan shows free contrast extravasation with gross

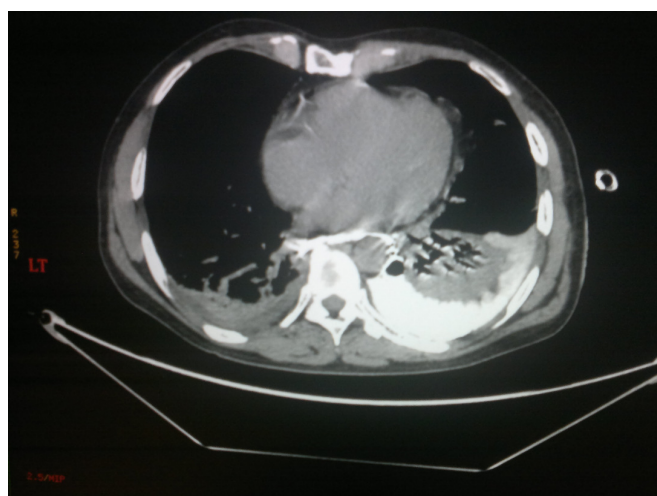


Figure 1: Contrast enhanced CT Scan of Chest showing B/L pleural effusion, Contrast extravasation in the pleural cavity and air pockets adjacent to distal esophagus.

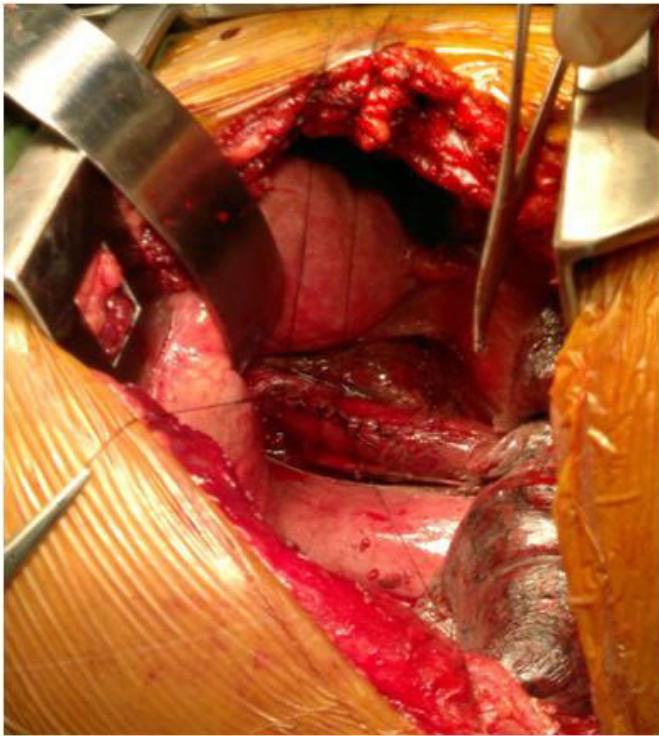


Figure 2: Perforation located in the distal esophagus being closed in layers.

mediastinal contamination (**Figure 1**) and if the patient is fit for surgery (**Figure 2**). A patient who is stable with CT scan showing minimal mediastinal collection without contrast extravasation is offered endoscopic stenting.

However if the patient is not fit for surgery then we offer endoscopic stenting in conjunction with intercostal drainage and feeding jejunostomy even when CT shows gross mediastinal contamination and free contrast extravasation with the idea that constant mediastinal soiling will be avoided.

Discussion

BS has mortality of 20-40%, highest amongst all causes of oesophageal perforation. Left untreated, it is invariably fatal.^{2,3} The high mortality is not only because of relative late detection in contrast to instrumental perforation but also because of release of acid, enzymatic juices and micro-organisms with high pressure leading to widespread mediastinal and pleural contamination.⁴

The presentation is often non specific, leading to diagnostic dilemma. Macklar's triad of chest pain, vomiting and subcutaneous emphysema may not be seen in all patients. It was seen in only one of our patients. It is not uncommon to see BS misdiagnosed as diaphragmatic hernia, myocardial infarction, or perforated duodenal ulceration before being

referred to a tertiary center for management.^{2,5} Three of our five patients were being treated as cases of pleural effusion before coming to us.

We have used multi slice helical CT scan of chest and upper abdomen with contrast for diagnosis. The findings in CECT suggestive of esophageal perforation are contrast extravasation, periesophageal air collection and pleural effusion. Published literature shows dominance of left pleural effusion but isolated right sided effusion is also noted, as seen in patient 3. Use of contrast esophagography may give false negative rates of 10% in detecting esophageal perforation.⁶

As the disease is rare no consensus exists on the treatment modality. All options, conservative, endoscopic and surgical management have been considered. In the absence of any level I evidence no clear cut management guidelines exist.²

Though most authors believe that surgery is the primary treatment modality, the type of surgery remains debatable. Options vary from primary repair to esophagectomy and cervical Esophagogastric anastomosis. Single stage Esophagectomy and cervical anastomosis is proposed as the first line of treatment in the setting of late presentation to prevent leak from suture line and resultant mediastinal sepsis. Esophageal diversion and exclusion is suitable for the cases with extensive devitalisation of oesophagus.^{4,7}

The traditional philosophy recommends primary repairs only within the first 24 hours. The success of primary repair in delayed cases (duration of perforation > 24 hours) is still not well established.⁸ Various authors have challenged this dogma and have reported excellent results with reinforced primary repair even in delayed cases.^{9,10,11,12}

Contrasting results from different authors perhaps call for meticulous technique for repair of these perforations.⁹ Thoracotomy on the side of perforation not only provides good access but is also useful in thorough mediastinal debridement and lavage.¹³ The mucosa and submucosa need to be entirely exposed by doing esophagomyotomy on either side of the perforation, this enables debridement of edematous and non-viable edges. Perforation can be closed in one or two layers. Reinforcement of repair with healthy tissue making sure not to cover the esophagus circumferentially decreases post-operative leak rates. A wide range of tissue e.g. pleura, diaphragm, pericardium, Intercostal muscle etc has been used by various authors to reinforce the repair. The chest is closed after full lung expansion.³ A feeding jejunostomy was done, so as to initiate early enteral feeding, this also ensures enteral nutrition in case of leak.

Minor leaks after primary repairs usually behave as controlled fistula. In the absence of mediastinal sepsis and with proper drainage, most heal without any further intervention and the outcome is not unsatisfactory.¹²

Endoscopic stenting for selected group of patients with minimum mediastinal contamination is an attractive option, which obviates the need for surgery.¹⁴

There is paucity of data regarding longterm outcome of these patients in literature. D' Journo et al reported mild GERD and nonspecific motor disorder in six out of seven of their patients on 24 hour pH monitoring and manometry.¹ None of our patients complained of dysphagia or increased frequency of reflux symptoms after a follow up of two years.

In conclusion, BS is difficult to diagnose and treat. A high index of suspicion is essential to diagnose it early as the outcome is dependent on the time lapse between perforation and institution of therapy. A multislice helical CT scan with oral and I/V contrast should be used to establish the diagnosis. We believe that acceptable morbidity and mortality rates can be achieved with primary reinforced repair. This along with the fact that it preserves the native esophagus and provides good quality of life in the long run makes it an attractive option, and should thus not be restricted to patients presenting within 24 hours, especially in a developing country setting like ours where the time gap between patients seeking medical advice and reaching a tertiary center is considerable.

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