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BOERHAAVE'S SYNDROME, OESOPHAGEAL PERFORATION, PRIMARY REPAIR

Medical Science

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ABSTRACT

Boerhaave's syndrome is a rare, potentially fatal condition. Diagnosis is difficult. Treatment is essentially surgical and success is based on a good post-operative management.We present a review of this condition along with a successful management of a 55 yr old female patient, diagnosed with Boerhaaves tear.

KEYWORDS

INTRODUCTION

Boerhaave's syndrome was first described in 1724 by Hermann Boerhaave, Professor of Medicine at Leiden University. In one of his publications in 1724, entitled "History of a Grievous Disease Not Previously Described", he discussed the case of Baron Jan van Wassenaer, a Holland Admiral, who after a feast had a big vomit followed by left side chest pain. The patient had no previous pre-existing illness. A tentative diagnosis of oesophageal rupture was made, although no treatment could be offered and the Admiral died. Autopsy revealed pleural fluid smelling of roast duck, which the baron had eaten the day before and a tear was in the transverse direction in the lower oesophagus[1].80% of the spontaneous or post emetic ruptures occurs in a normal esophagus; only 10% have a previous history compatible with esophagitis or ulcer. Sensitivity of the esophageal mucosa to gastric juice weakens the wall and allows perforations to occur with less pressure. However, this does not prove that all spontaneous ruptures are due to esophagitis. To complicate matters, esophagitis is hard to recognize at postmortem examination since an inflammatory reaction occurs around the disruption. The importance of persistent retching leads one to suspect that spontaneous rupture occur on an empty stomach. Several spontaneous perforations have been reported in newborn infants. Esophagomalacia secondary to central nervous system disease contributes to a type of spontaneous rupture[2].

Epidemiology

Boerhaave's syndrome accounts for approximately 15% of all cases of esophageal rupture. The incidence worldwide is unclear, estimated at approximately 3.1 per 1,000,000 per year. The patient population who suffer from Boerhaave's syndrome varies, with the highest risk in middle-aged (40-60%) males (80%)[3].

Pathophysiology

Boerhaave's syndrome is due to a sudden rise in the intraluminal esophageal pressureagainst a closed glottis.Typically, the adult history includes food or alcohol overindulgence and vomiting. There is failure of the cricopharyngeous muscle to relax due to neuromuscular incoordination. The sequelae following rupture will depend on the location of tear, as the esophagus abuts different areas of the body. The mid esophagus lies next to the right pleura while the lower esophagus abuts the left pleura. Rupture occurs most commonly in the left poster lateral wall of the distal third of the esophagus. The rent almost always occurs in the lower third of the esophagus on the left posterolateral wallwith extension into the left pleural cavity.The tear is usually linear, averaging 2.5 cm. in length. The thoracic aorta and retropericardial fat support the esophagus on three sides, leaving the least supported lower left side to be most affected. The musculature at the lower end of the thoracic esophagus is thin and terminates in a conical fashion, segmental defects are found in the circular muscle at the distal end; and the wall is further weakened by the entrance of vessels and nerves in this region. The bursting pressure ranges from 3.5 to 6 pounds per square inch and creates a longitudinal defect. Mucosa bulges through the weakened area and offers a greater resistance than the muscular layer[4].Another site of rupture is sub-diaphragmatic, leading to abdominal symptoms, however the liver and gastric fundus stabilize the short segment of abdominal esophagus and along with the peritoneum and phrenico—esophageal fascia makes this area fairly strong. The cervical ruptures are usually more localized and benign as the spread to the mediastinum through the retroesophageal space is limited and slow when integrity of the thoracic esophagus is lost,mediastinal contamination occurs, in which case, saliva and gastric contents create a fulminating chemical and bacterial effect in the mediastinal tissue, the principal feature of which is hemorrhagic necrosis. The thin mediastinal pleura quickly rupture from continuous soilage, allowing negative intrathoracic pressure to draw gastric fluid through the perforation. The subsequent pleural space contamination produces more fluid loss that soon leads to hypovolemia and shock. Fluid aspirated from the thorax is highly contaminated by food and assumes a brownish color as blood is acted on by the gastric juice. Pleural fluid may accumulate as rapidly as 1,000 ml. per hour, compressing the lung, shifting the mediastinum, and further embarrassing the cardiorespiratory mechanism. The increased pressure in one or both thoracic cavities eventually retards venous return. If the perforation point is very small in the middle thoracic esophagus, and the wall of the esophagus is thickened circumferentially along its entire length, it represents chronic diverticulitis or esophagitis. In addition, the squamous epithelium is interposed in the esophageal depression pathologically, providing evidence of the presence of an esophageal diverticulumIntrathoracic esophagus perforations lead to mediastinal inflammation and necrosis due to gastric contents entering the area[6]. Pre-existing oesophageal disease delays the diagnosis. Perforation in esophageal diverticula is relatively rare and was reported by Mellius at an incidence of 0.15% (23 cases of over 15,000 consecutive autopsies). Stasis in chronic diverticulitis with bacterial overgrowth causes localized inflammation and ulceration result in perforation. Ifdiverticula are noticed at the time of surgery, a procedure to address the distal obstruction like a distal myotomy is contemplated at the time of primary surgery[7].

DIAGNOSIS ofspontaneous ESOPHAGEAL PERFORATION
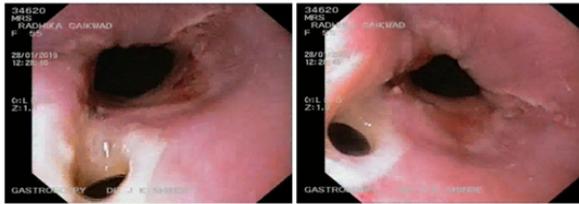
The clinical presentation of Boerhaave syndrome will depend on the site of the perforation, the degree of leakage, and the time elapse since onset. Pain is the striking symptom, searing or bursting sensation at the time of perforation in the epigastric or substernal region, later extending laterally to the lower thorax assuming a pleuritic character. The typical history of overeating and alcoholic debauch are few features. Vomiting, if present, subsides as the pain increases. The patient appears pale, sweating, occasionally cyanotic, and frequently dyspneic, and unless in profound shock, is febrile. Hematemesis is seldom significant-a means of differentiating perforation from the Mallory-Weiss syndrome. Mediastinal air soon tracks upward, and subcutaneous emphysema is palpable in approximately 60% of the cases. The symptoms and signs may be accentuated by deep breathing, swallowing, or changes in position.

The single most valuable diagnostic procedure is the upright chest roentgenogram. The majority of patients have pleural effusion or hydropneumothorax, and 7% have bilateral effusion. X-Ray reveals air in the fascial planes of the mediastinum, particularly in the retrocardiac area. Later the mediastinum widens and air can be seen in the soft tissues of the neck. The presence of subdiaphragmatic air is extremely rare. If the roentgenogram is underexposed or taken too early, mediastinal emphysema may be missed.

Controversy exists over the type of contrast material to be employed foresophagograms. Barium acts as a foreign body in an infected area when it extravasates in the pleural cavityleading to mediastinitis with subsequent fibrosis. Others believe that the contrast produced by barium is far superior to that of iodinated contrast media, and point out that barium can be removed by irrigation during operative treatment of the defect. Lack of extravasation of the radiopaque medium does not necessarily exclude the presence of a perforation. Passage of the material may have been too rapid for the small size of the rent, or its location may have been viewed in a projection parallel to the body of the esophagus; thus the leak would be overlooked.

When the esophagogram is normal and doubt concerning the diagnosis persists,CT scan is used by due to its higher sensitivity and more detailed assessment of the involved organs.

Carter and Hinshaw recommendesophagoscopy as a diagnostic procedure[8].Endoscopy is useful but done with caution due to the risk of further aggravating the injury and increasing the contamination because of insufflation. Endoscopy can diagnose a perforation, but may not be able to focus the cause which could vary from a spontaneous rupture toa perforated diverticula.Since surgery has to be done early, it may not be possible to further evaluate the case. If a chest tube is present, one can also use the methylene blue dye test. When sweetened methylene blue is taken orally, it gives a bluish discolouration to the chest tube effluent within 12 – 24 hours.

Differential diagnosis is acute pancreatitis. The serum amylase content and roentgenograms of the chest demonstrating mediastinal or subcutaneous emphysema are the best aids in distinguishing the two. Electrocardiogram and chest findings help exclude myocardial infarction. Pain from a dissecting aneurysm peaks during the initial dissection and subsides thereafter. Spontaneous pneumothorax does not produce the severe pain or shock that occurs with perforated esophagus and pleural effusion develops as a late result. Mesenteric thrombosis may occur in elderly patients with a recent myocardial infarction or arrhythmia; however, the abdominal pain does not change in location and often is unrelenting despite narcotics. Other conditions to consider include pulmonary embolism, intraabdominal abscess, strangulated diaphragmatic hernia, and acute cholecystitis[9].One-third of patients have atypical symptoms or are admitted with severe respiratory distress and/or shock. Physical exam findings may include abnormal vitals (tachycardia, tachypnea, fever); decreased breath sounds on the perforated side, mediastinal emphysema, and Haman's sign (mediastinal "crackling" accompanying every heart beat) in left lateral decubitus position. The syndrome of vomiting, lower thoracic pain, and subcutaneous emphysema (Maklers triad) isseen in 33% of cases.

Treatment / Management

Treatment is tailored to the clinical presentation, type and extent of rupture, time to diagnosis, and the viability of the esophageal wall. Early perforations, diagnosed within 12-24 hours, have the best outcomes. Management of Boerhaave's syndrome is a controversial issue. Three levels of treatment are distinguished: conservative, endoscopic, and surgical [10]

Conservative measurements are reserved for small and contained ruptures. There are three circumstances in which this management play a limited role: (1)delayed diagnosis, (2) gravely ill patient, and (3)rupture of the thoracic esophagus. Barrett's statement summarizes the current attitude toward conservative management: ". . . to treat a perforation conservatively may succeed; but it is likely to fail in that the patient who survives the emergency develops a mediastinal abscess or pleural complications and these result in prolonged illness and pathologic sequelae which can be virtually untreatable." . The least publicized aspect is the prolonged disability and hospitalization which accompany this choice of treatment.Conservative treatment consists of cessation of oral intake, administration of I.V.fluids with parenteral nutrition, broad-spectrum antibiotics, H2-blockers, and eventual mediastinal, pleural, or abscess drainage. Parenteral hyperalimentation is an useful adjuvant therapy[11].

Endoscopic placement of stents have been used to prevent fistula formations or seal esophageal leaks in both patients with delayed diagnoses and those with the early diagnosis without widespread contamination. Endoluminal placement of a self-expandable metallic stent to bridge an esophageal tear has shown encouraging results. Endoscopic stenting in Boerhaave's syndrome has an undetermined and center-dependent success rate and may not be devoid of side effects such as enhanced mediastinal or pleural contamination and accidental stent migration[12]
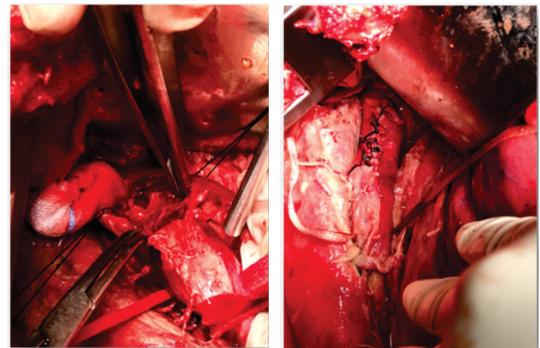
Surgery is the radical sanction. Factors determining surgical intervention are the extent of the perforation. Surgical intervention include primary esophageal repair through open thoracotomy vs. VATS with fundic reinforcement. Controversy occurs when presented with a late perforation, as the wound edges becomeedematousand friable rendering primary repair risky. Taking this into consideration, late perforations are managed through debridement of pleural cavity and mediastinum, esophagostomy, and feeding gastrostomy. Esophageal replacement is usually done after 6 weeks[13].

Abbas and colleagues report the mortality of thoracic perforation that have undergone repair as 38.5%. Wright and colleagues report no deaths when treatment was initiated within the first 24 hrs, but the mortality rate was 31% when treatment was initiated after 24 h, and the delay in diagnosis and treatment significantly influenced mortality[14].Experience with less invasive surgical procedures is scarce. Haveman et al. compared open thoracotomy with video-assisted thoracoscopic surgery (VATS) and found similar success rates. More prospective studies are warranted to establish the potential advantage of VATS in treating patients with Boerhaave'ssyndrome[15].

Collis and his associates accurately diagnosed a spontaneous rupture in 1944 and were the first to perform a thoracotomy for closure of the rent; unfortunately, the patient died.Barrett's review of spontaneous perforation two years laterpredicted the success of direct surgical repair, provided that the diagnosis could be made early. Shock is not a contraindication to operative treatment. A standardleft postero lateral incision in the seventh intercostal space is taken.Despite a layered closure and tube decompression, breakdown of the repair may occur because of edema or necrosis of the suture line. When the underlying process is esophagitis, this alone may be responsible for poor healing. Inadequate drainage results in empyema, atelectasis, and continued mediastinitis. Early postoperative course suture-line dehiscence produces increase the drainage through the chest tube. Late disruptions create roentgenographic evidence of an air-fluid level and a febrile course. Postoperative breakdown of the repair does not obligate the surgeon to reoperate for a second closure, provided that the leak is well drained. These fistulas close when the patient's nutrition is maintained and there is no obstruction below the leak.

The standard operative techniques must be modified when lower obstruction underlies the perforation. Thal'sfundic patch operation is applied to perforations with stricture. The procedure was designed to widen a narrow lumen and provide a valve mechanism at the cardioesophageal junction[16].In critical cases of mediastinitis, Johnson and Schwegman's method of esophageal exclusion may be lifesaving; however, it does entail secondary reconstruction of the esophagus. A cervical esophagostomy is created and the lower esophagus is divided in the abdomen. The proximal and distal ends of the esophagus are closed and the pleural cavity is drained. A gastrostomy is used for feeding. Later, the blind esophageal segment can be removed and the colon transplanted to restore continuity.

Our case with a successful outcome

Medical History
55yr old female was admitted for severe chest pain and dyspnea offollowing a bout of vomiting after food accompanied with mild hematemesis. There was history of viral fever 7 days prior to this and patient had cough and had taken treatment for 2 days for the ailment. X-Ray chest and CT scanshowed bilateral basal pneumonia with pleural effusion. A tentative diagnosis of viral pneumonia with? Mallory Weis tear was made.

Gastroscopy done showed a perforation in the left lateral wall of esophagus.About 2cm above OG junction. The perforationwas circular in shape, 1cm in diameter with no ragged margin and the mucosa inverting into the opening. With these findings a possibility of a perforated diverticulum was contemplated. There were no visible signs of distal obstruction. Patient taken up for surgery.Chest opened via left, 7th rib excision incision.The exploration showed a purulent collectionwith extensive fibrinous adhesions.A perforation 2-3 cm from the oesophageal hiatus was foundthe tear was longitudinal with ragged margins and mucosa everting from the torn muscularis. There were no diverticula. The tear was freshened till the mucosa adequately pouted from the tear. The repair of the esophaguswasperformedin two

layers with interrupted sutures with 2'0'ethibond. Sutures were full thickness with a good bite of mucosa. Since the repair seemed sound without tension, fundic overlap was not considered. Pleural cavity toilet was performed. Anterior and posterior chest tubes drain were placed inthe pleural space. Thorax closedin layers. Fluid in the opposite hemithorax required insertion of a chest tube on the right side.A feeding jejunostomy was performed.Longitudinal oesophagealmyotomy is done as the mucosal injury is usually longer than the muscular one and mucosal debridement may additionally be necessary.Post-operative is the routine intercostal drain management,Ryle's tube aspiration along with supplementary parenteral hyperalimentationstarting from the 1st day followed by enteral nutrition later. Post-operative period was uneventful.Post-operative swallow done on 15th day showed a minimal leak with no distal obstruction. Patient was started on oral fluids. After a week, a repeat swallow showed no leak and patient started on regular feeds and discharged after 2 days.Patient has been following for 6 months with an uneventful period.

Leak rate after primary repair performed within the first 24 hours of presentation is in the range of 20% and if treatment is delayed for more than 24 hours it rises to 50%.

Pre-operative a suspicion of perforated diverticula was considered because on gastroscopy the perforation was circular with no ragged margins. In addition there was no mucosal tear which seemed to pout outwards. Esophageal diverticula are rare, with a prevalence of 0.06% to 3.6% based on radiologic and endoscopic series[17].

CONCLUSION
Diagnosis of Boerhaaves syndrome is challenging. Management should be prompt and multidisciplinary,which reduces mortality. Some centers advocate early and extensive surgery, yet others prefer a more conservative endoscopic, or less invasive surgical approach. Such differences explain the lack of well-established treatment guidelines. The key is maintaining vigilance and suspicion of the diagnosis when assessing both medical and surgical emergency admissions.Early surgery is the path for a successful outcome.

The high risk of developing malignancy has also been described, for which the carcinogenetic factors include chronic mechanical irritation by food, repeated physical injury and inflammation; however, the involved mechanism remains elusive. Esophageal diverticula are classically divided into two types: pulsion (false) and traction (true) diverticula. Pulsion diverticula occur as a result of increased intraluminal pressure and form protrusions of mucosa through the muscular wall; therefore, they lack muscularispropria. Zenker's diverticulum and epiphrenic diverticulum are included in pulsion diverticula. Traction diverticula are created by the pulling force of the outside esophagus due to periesophageal inflammation, fibrosis and adhesions to surrounding structures. Most traction diverticula occur in the mid-esophagus, and the common causes include tuberculosis, histoplasmosis and malignancy. Most diverticula are recognized as mucosal depressions on endoscopy.[18].However the patient did not have perforated diverticula on the operation table and the findings were in favour of a spontaneous rupture.

The overall mean time to theatre was 2.4 days from symptom onset. It was higher in those patients who did not survive.19.

Chronic morbidity is common after Boerhaave's syndrome and 30-40% of surviving patients in this series went on to experience further oesophageal morbidity after discharge.

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