Survival after distal esophageal rupture: A case of Mallory Weiss tear becoming Boerhaave's Syndrome during a pandemic.

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INTRODUCTION

We present the case of a 42 year old male cirrhotic chronic alcoholic who was admitted from home in April 2020 during the height of the COVID pandemic for altered mental status, history of upper GI bleeding due to retching, agonal respirations with large right pleural effusion. Initially 2 liters of dishwater colored fluid was drained after placement of a pigtail thoracostomy tube, but the patient remained too unstable for procedural interventions and over the course of three weeks he was resuscitated in critical condition in the ICU. Upon downgrade to the medical floors and initiation of oral liquid diet, he was observed to have output of liquid diet in his chest drainage system. Further investigation revealed a large distal esophageal rupture on the right side near the gastroesophageal junction as the source of his chronic pleural effusion and he was diagnosed with Mallory Weiss tear converted to Boerhaave's syndrome. He underwent successful covered stent placement with gastroenterology, but had a protracted course with presumed empyema secondary to gastric contents in his chest. Eventually the patient was taken for thoracotomy with decortication and wash out, he recovered well post-operatively and was discharged home on an oral diet. Our case provides an excellent example of distraction from a global pandemic secondary to similar presenting symptoms of COVID, and a prolonged critical condition preventing invasive procedures, leading to significant delay in recognition of an esophageal rupture. However, he is uniquely impressive when compared to similarly published cases of Mallory Weiss conversion to Boerhaave's Syndrome, given his significant increased risk of morbidity based on his disease course and his survival with excellent clinical outcome leading to discharge home on oral diet.

CASE DESCRIPTION

We present the case of a 42 year old male with history of alcohol and cocaine abuse who was brought to the emergency department from home in April 2020 at the height of the COVID-19 pandemic in the New York City area. As per his family he complained of hematemesis leading to shortness of breath and eventually he became unresponsive leading to their emergency call. In the emergency department, our patient was persistently tachycardic and required increasing supplemental oxygen therapy, requiring admission to the intensive care unit (ICU) as a person under investigation for COVID-19. No episodes of hematemesis or melena were noted, he had stable hemoglobin and hematocrit, however due to his recent history of hematemesis, gastroenterology was consulted. Endoscopy was deferred given patients clinically tenuous status and lack of obvious GI bleeding. On hospital day 3, the patient's respiratory status continued to deteriorate and a portable chest x-ray demonstrated a large right pleural effusion prompting general surgery consultation for thoracentesis and chest tube placement. Upon evaluation by the surgical team, our patient appeared toxic with acute respiratory distress. He was saturating 90-92% on 15L oxygen via nonrebreather, tachypneic to 40 breaths per minute, using accessory muscles of respiration, and breath sounds on the right were severely decreased. Additionally, on physical examination he had a distended abdomen with fluid wave consistent with ascites. Laboratory examination demonstrated Child-Pugh Class B cirrhosis, leukocytosis of 20,000 white blood cells/mm^3, and acute versus chronic kidney injury with a creatinine of 2.3 mg/dL. Informed consent was obtained from our patient's spouse for tube thoracostomy and a 14F pigtail catheter was placed in the right 5th intercostal space anterior axillary line. Immediately 2 liters of dirty dishwater colored fluid was drained and the chest tube was then clamped to decrease risk of flash pulmonary edema and unclamped several hours later.

Over the following 2 days, our patient was determined to be COVID-19 negative, clinically improved, and was downgraded to the medical floor. On hospital day 6, he became febrile and more tachycardic; However, his oxygen requirements had decreased to 2-4L nasal canula titrated to maintain oxygen saturation above 94%. Non-contrast computed tomography of his chest demonstrated a continued right sided pleural effusion with multi-loculated fluid collections with air-fluid levels around the thoracostomy tube. Cardiothoracic (CT) surgery was consulted for management of the visualized loculated pleural effusion. At this time, it was determined that the patient was not a surgical candidate as his clinical status would not tolerate single lung intubation or surgical stress. The pigtail thoracostomy tube was continued to a chest drainage system on suction and antibiotics were continued with a regimen tailored by infectious disease for pleural cultures which eventually speciated to *Candida Albicans* which was also observed in the patient's sputum. The next day, our patient returned to the ICU on a required Ativan drip due to worsening alcohol withdrawal which progressed to delirium tremens. His thoracostomy tube remained on suction but subsequent chest x-rays did not show improvement of the effusion despite changes in antibiotic regimen with antifungal coverage. On hospital day 9, he underwent interventional radiology placement of two new image guided pigtail thoracostomy tubes into the loculations noted within the right pleural effusion, and the prior surgically placed thoracostomy tube was removed (Figure 1). These new pigtail catheters initially drained 300mL of purulent fluid superiorly, and 50mL of purulent drainage inferiorly. He also underwent peripherally inserted central venous catheter (PICC) insertion for total parental nutrition (TPN).

After aggressive medical management, antibiotics, and recovery from alcohol withdrawl, our patient was once again downgraded to the medical floors. His pigtail thoracostomy tubes had begun to decrease in output, however he had persistent effusion on portable imaging prompting further evaluation in interventional radiology. Both pigtail catheters were upsized for 28F chest tubes resulting in more adequate drainage of his loculated pleural effusion (Figure 2). On hospital day 17, his medical conditions and mental status had improved enough for the medical team to begin oral feedings with clear liquid diet. Several days later, he was observed to have bright green output in his chest tube drainage system after eating green Jell-O. He was immediately made nil per os (NPO) by the medical team pending repeat evaluation by CT surgery. Upon surgical evaluation, his chest tube output was noted to be clear and foamy rather than purulent or straw colored, and raised concern for salivary contents as opposed to a traditional transudative effusion. At this time the surgical team also noted that he had regained normal mental status and he was updated on the events over the past three weeks of his hospital stay. He recalled events leading up to his hospitalization and provided a more detailed history of the events leading up to his hospitalization. He indicated that he had been binge drinking beer and red wine the weekend prior to his admission, but was unable to keep any solid or liquid intake down due to forceful retching. Despite retching, he admitted to continuation of his alcohol consumption and intra-nasal use of cocaine over the course of three days. He was unable to verify hematemesis as he indicated that the wine he was drinking was red and that his emesis was the color of the consumed wine. He reported his final memory prior to this day was that of severe shortness of breath and blacking out in his bedroom with his spouse attempting to revive him but he was unable to respond.

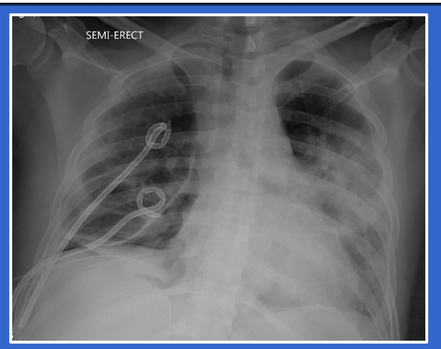

Figure 1. Anterior-Posterior (AP) chest x-ray demonstrating two 14F pigtail thoracostomy tubes in right hemithorax.

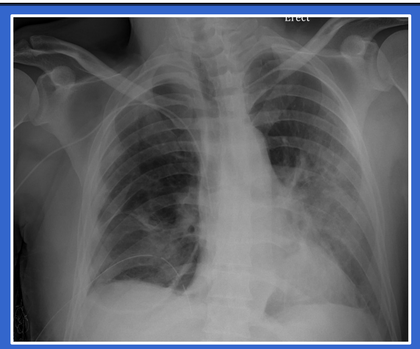

Figure 2. AP chest x-ray demonstrating upsizing to two 28F thoracostomy tubes in right hemithorax.

At the time of repeat CT surgical evaluation, our patient's vitals had stabilized and he was no longer febrile or tachycardic. He requried minimal to no supplemental oxygen and had non-labored respirations on room air when examined by our team. At that time, our differential diagnosis included chronic aspiration with empyema and lung abscess secondary to chronic alcohol intoxication, or an esophago-pleural fistula secondary to esophageal disruption from forceful retching. A gastrograffin swallow evaluation was ordered and performed the following day demonstrating a distal esophageal perforation with preferential flow of contrast into the right pleural cavity (Figure 3).

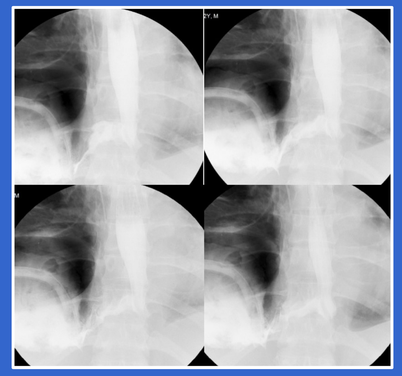

Figure 3. Gastrograffin esophagram demonstrating distal esophageal perforation with flow into right pleural cavity.

Figure 4. Calcified object and three metallic endoscopic clips removed from right hemithorax during thoracotomy.

After extensive conversations with our patient and his wife, he was consented for a planned surgical cervical spit fistula and the general surgery team was consulted for feeding jejunostomy tube placement. He also understood that he may additionally require thoracotomy and decortication for empyema, but that we were attempting to control his infection and drainage with the chest tubes given the chronicity of his esophageal perforation at the time of diagnosis. After general surgical team discussion with the patient and the gastroenterology team, the planned spit fistula was subsequently canceled to allow endoscopic covered stent placement over the perforation in addition to a percutaneous endoscopic gastrostomy (PEG) tube placement by gastroenterology. After this procedure was completed the medical team began PEG tube feedings, leading to a notable increase in chest tube output, prompting return to the original CT surgical recommendations for feeding jejunostomy tube. The PEG tube was converted to a feeding jejunostomy tube by interventional radiology and this was utilized for enteral feeding.

A repeat esophagram was performed demonstrating a small amount of contrast leak through the esophagus external to the stent into the right pleural space. Esophageal reflux was also noted above the level of the stent. Computed tomography of the chest with oral contrast demonstrated improvement of the empyema with a small persistent effusion and loculated pocket of air. Jejunostomy tube feedings at that time were also associated with increased chest tube output, concerning for continued esophageal leak. Repeat fluid analysis at this time revealed fluid with a pH of 7.5, amylase of 18,477 u/L, total protein of 3.7 g/dL, red blood cell count of 569,250 cells/mm^3, and white blood cell count of 85,770 cells/mm^3 with a differential of 94% neutrophils. Fluid lipid analysis and chylomicrons were unable to be obtained due to laboratory capabilities. Cultures demonstrated moderate coagulase negative *Staphylococcus* and *Stenotrophomonas maltophilia*, and *Klebsiella oxytocci*. CT surgery team recommended a venting gastrostomy tube to decrease pressure on the gastroesophageal junction and distal esophagus from swallowed oral contents and refluxing jejunostomy feeds. This venting PEG tube was placed by gastroenterology several days later.

Our patient continued to improve and was eventually advanced by the medical team to clear liquid diet which he tolerated well with intermittent requirement for venting from the gastrostomy tube. A repeat chest computed tomography demonstrated loculated collection of air regional to chest tube within lower right hemithorax with radiodense structures in the dependent aspect of this space. A repeat barium esophagram demonstrated no leak or aspiration. Chest tube output remained minimal and our patient was continuing to tolerate clear liquid diet; therefore, supplemental jejunostomy tube feedings were stopped. On hospital day 54, the remaining chest tube was removed which the patient tolerated well without pneumothorax or significant re-accumulation of effusion post procedure. Chest computed tomography with oral contrast the following day demonstrated a very small leak of contrast between the esophageal wall and the stent at the site of previous perforation and a small area of round atelectasis in the right hemithorax anterior to a small amount of pleural fluid. CT surgery recommended that the patient return to NPO status with TPN for 3 to 4 weeks further to allow for full healing of the perforation. However, he was discharged by the medical team several days later with instructions to advance diet as tolerated.

After one week at home on regular diet, our patient returned to the hospital complaining of burning post-prandial chest pain. His admission labs revealed significant leukocytosis of 17,500 cells/mm^3, and imaging demonstrated an increased right pleural effusion with a 10.8cm by 6.2cm area of loculated air and fluid collection in the posterior right hemithorax. Gastrograffin esophagram demonstrated no evidence of esophageal leak, and interventional radiology was consulted but deferred chest tube placement due to heavily loculated effusion unlikely amenable to drainage with percutaneous drainage alone. CT Surgery was re-consulted and after discussion with the patient and his wife, he was consented for right chest video assisted thoracoscopy (VATS) and possible open thoracotomy with decortication for chronic empyema due to chronic effusion of gastric contents. He was brought to the operating room where VATS was attempted but quickly aborted secondary to inability to achieve visualization due to severe adhesions in the pleural space. We converted to open thoracotomy, which required a single rib resection to obtain adequate visualization and room for adhesiolysis. Two large areas of loculated effusions were discovered with gross purulent empyema in the posterior inferior loculation. This purulent material was suctioned and sent for culture which eventually grew *Klebsiella oxytoca*, *Enterococcus faecalis* group D, and few *Candida albicans*. A 0.5cm calcified object was removed from this cavity along with three metallic clips from previous endoscopic management (Figure 4). Two large bore chest tubes were left in each loculated pocket and a wound vac was applied over closed fascia and widely spaced interrupted skin closure to prevent post-operative soft tissue infection. Final pathologic reports demonstrated an area of acute osteomyelitis in the resected rib, amorphous calcific material with scattered neutrophils and macrophages, metallic foreign bodies thought to be esophageal clips from previous endoscopic treatment of the esophageal tear.

The postoperative course was complicated with development of acute kidney injury which was managed with intravenous fluid hydration by nephrology. Our patient was transitioned from parenteral nutrition to enteral feeding via jejunostomy tube with the gastrostomy tube clamped. At this time, he developed nausea and persistent emesis with any rate of feeding over 10mL per hour. The gastrostomy tube was replaced to venting suction and jejunostomy tube feedings were held. The chest tube output was also noted to be thick and milky white in consistency, raising concern for chylothorax after initiation of a diet containing lipids. Our patient was slowly advanced to clear liquid diet with TPN supplementation without intralipids. His chest tube output cleared up over the next several days and he continued to improve clinically. The chest tubes and wound vac were removed based on patient progression and his labs eventually normalized. He was discharged home on low fat, medium chain triglyceride clear liquid diet and has subsequently been advanced to a more substantial diet without any complications.

DISCUSSION

Spontaneous esophageal rupture (Boerhaave's syndrome) can manifest as an array of symptoms including classic Mackler's triad of vomiting, lower thoracic pain, and subcutaneous emphysema,[1,2] as well as less common symptoms including dysphagia, tachycardia, fever, tachypnea, and epigastric pain.[3] These perforations are usually longitudinal, average 22mm in length, and are most common approximately 3-4cm above the gastroesophageal junction.[1-3] Furthermore, esophageal rupture is most common on the left side due to anatomical weakness of the left posterolateral aspect of the esophagus just above the diaphragm, resulting in full thickness rupture of the esophagus.[1-3] Whereas, Mallory-Weiss tears are more likely to occur on the right side of the esophagus.[3] It is thought that only 50% of Boerhaave's syndrome present with Mackler's triad which contributes to a significant delay in diagnosis for many patients, and the historical treatment option for prevention of fatality in this rare condition has been prompt diagnosis with thoracic drainage and damage control via surgical approach.[1] Other cases have documented delayed presentation due to misdiagnosis initially and recommend that Boerhaave's syndrome should always be considered in all ill appearing patients presenting with a combination of gastrointestinal and respiratory complaints.[4] Principles of management are the same for all patients, including sepsis control by limiting diffuse contamination, adequate drainage, perforation repair and antibiotic therapy to prevent the most common causes of death from sepsis and multiple organ failure.[1,4]

Currently, only three publications report conversion of Mallory-Weiss tear to Boerhaave's syndrome over the course of 50 years. McVay et al. first reported a case of conversion in 1970, and no further reports were made of this phenomenon until 2002 when Marshall documented another case from the anesthesia perspective after 15 days of follow-up in the intensive care unit.[5,6] In 2018, Cucci et al. reported a case of a 45 year old male who unfortunately succumbed to cardiorespiratory arrest due to pleural effusion after spontaneous transition from Mallory-Weiss lesion to Boerhaave's syndrome over a few days. Diagnosis greater than 24 hours from the onset of symptoms has been proven to increase risk for fatal outcome.[3,7]

A retrospective review of 34 patients with spontaneous esophageal rupture over 30 years, found that 50% of patients had a delay in diagnosis defined as time from onset of symptoms to establishment of correct diagnosis exceeding 24 hours often secondary to patient delay and incorrect initial diagnosis, with 77% resulting from diagnostic delay as the most common cause. This review did determine that the delay in diagnosis did not affect mortality rates, with mortality of 33% in those patients diagnosed within the first 24 hours and 34% in those diagnosed after 24 hours. They stressed that broad-spectrum antibiotics for treatment of mediastinitis, nasogastric decompression, sump drainage of the upper esophagus, antibiotic fluids by mouth, correction of fluid and electrolytes as well as blood volumes before during and after any operative intervention are key to this result.[8] Five years later, Troum et al. reported the first case of surviving Boerhaave's syndrome with free pleural rupture without requiring thoracotomy in literature in 1994.[9] Later, in 1996, Jagminas et al. reported a case with delayed diagnosis of Boerhaave's syndrome with right sided esophageal perforation due to unusual symptoms on initial presentation.[4]

More recently, information has been published detailing non-surgical management of Boerhaave's syndrome with a two year follow-up observed.[2] There were no deaths in this study group and clinical resolution was achieved in all patients without any subsequent surgical intervention. However, the mortality rate observed in this small case series was 30-50% in patients who had a delay in diagnosis. The most common site for perforation was the distal esophagus, 3cm above the gastroesophageal junction. 83.3% of patients required mechanical ventilation for respiratory failure. 100% of the patients had endoscopic treatment with full covered esophageal stents and had interventional radiological (IR) drainage of mediastinal and pleural fluid. 50% of these patients required re-stenting due to stent migration or continuous leakage at the perforation. 66.6% of the patients had subsequent feeding tube placement via gastrostomy or jejunostomy for enteral nutrition until complete resolution without surgical intervention. They also report that Boerhaave's diagnosis is often delayed due to vague clinical picture when compared to iatrogenic rupture, thus leading to detrimental effects on survival. This recent information proposes that esophageal stenting is sufficient management of complete esophageal rupture to prevent continued septic contamination and guides re-epithelialization of esophageal mucosa with stent placement or endoscopic suture. The authors propose that closure of the mucosal defect also allows for early enteral feeding and a great reduction in morbidity and mortality when compared to surgical management. However, the main drawbacks of esophageal stenting alone include migration, pressure-induced ischemia, ulceration, and perforation, new reactive stenosis at the ends of the endo-prosthesis, bleeding or injury upon removal of stent, and unsuccessful retrieval of device later.[2] We also discovered in our patient that misplacement of endoscopic clips through the perforation into the chest creates a significant risk for morbidity and should not be overlooked in patients with ongoing empyema or pleural effusions due to foreign body reaction.

In the past, nonoperative management of esophageal perforations was only considered for circumscribed transmural perforation, non-neoplastic tissue, not extending into the abdominal cavity, not accompanied by simultaneous obstructive esophageal disease, and cannot have any evidence of septicemia.[10] A retrospective review of all options for management of all esophageal perforations in 44 patients over 12 years found that 27% of the patients underwent esophageal endoscopic stenting, 55% of the patients underwent surgical management, and 18% underwent conservative treatment with cessation of oral intake, antibiotics, and parenteral nutrition. Of the surgically managed patients, 20% required suture closure of the lesion, 32% required esophagectomy with delayed reconstruction, and 1% required distal esophageal resection with gastrectomy. Overall this retrospective review observed that 89% of patients with Boerhaave's syndrome underwent surgical management.[11] More recently, Schweigert et al. compared outcomes for endoscopic stent insertion versus primary operative management for Boerhaave's syndrome.[7] 84.6% of patients who underwent endoscopic stenting developed either severe pleural empyema or sustained mediastinal abscess formation, similar to our patient's clinical course, and eventually required operative intervention via video assisted thoracoscopic surgery, thoracotomy, or mediastinotomy, and 23% of these patients required repeated surgical intervention. Two patients in this group required open thoracotomy with decortication of the lung. This study is unique because it only evaluated patients with spontaneous rupture due to Boerhaave's syndrome and eliminated data from esophageal perforations that resulted from iatrogenic sources or from esophageal cancer. Although surgical intervention was nessary to manage infectious complications from gastric contents in the chest, death rates were not statistically significant between the stent group and surgical group.[7]

CONCLUSIONS

Boerhaave's syndrome should be considered in all ill appearing patients presenting with the combination of gastrointestinal and respiratory complaints. Unfortunately, in our patient's presentation, his history of forceful wretching was not obtained until after he was stabilized and extubated due to his initial presentation to the emergency department in severe respiratory distress and non-responsive state (4). Nonoperative management of esophageal perforations historically was only considered for circumscribed transmural perforation, non-neoplastic tissue, not extending into the abdominal cavity, not accompanied by simultaneous obstructive esophageal disease, and cannot have any evidence of septicemia.[10] However, more recently, it has been shown that a delay in diagnosis of Boerhaave's syndrome is often due to a vague clinical picture when compared to iatrogenic rupture, and a delay more than 24 hours increases risk of fatal outcome overall but without statistically significant differences in patients treated with stents versus surgically.[2,7,8] Our patient is uniquely impressive when compared to similarly published cases of Mallory Weiss conversion to Boerhaave's syndrome, given his significant increased risk of morbidity based on his disease course and his survival with excellent clinical outcome leading to discharge home on oral diet.

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