Case Report

Open Access

A rare case of sub-diaphragmatic oesophageal perforation resulting in peritonitis

Abstract

The case under discussion elaborates a death following full-thickness lower oesophageal perforation resulting in generalized peritonitis. Causes for oesophageal perforation are many including Boerhaave syndrome. A 14 year old mentally subnormal girl with a past history of epilepsy became acutely ill with fever, vomiting and diarrhoea for three days. She died soon after hospital admission without much room for investigations or interventions. No preliminary resuscitative attempts such as intubations were on record. An inquest was requested and a post-mortem examination was performed. A longitudinal perforation measuring 7x2.5 cm with necrotic borders was identified in the lower oesophagus. There was no evidence of trauma or any other disease that could be attributed to the perforation. Generalized peritonitis due to lower oesophageal perforation was stated as the cause of death although the underlying cause for the perforation was undetermined. Post mortem oesophagomalasia is a rare but recognized entity. This may be mistaken for an antemortem perforation by an inexperienced medical officer. Even rarely an antemortem perforation may be mistaken as post mortem oesophagomalasia. Though a brief summary of the autopsy findings is included in the bed head ticket after a post mortem examination, it rarely results in initiating a clinico-pathologic dialogue. It is strongly recommended that clinicians pay a renowned interest on the autopsy findings especially if the cause of death is not established in the ward. This may make the clinicians aware of the rare and unsuspected clinical entities which may prove to be fatal at times.

Keywords: oesophageal perforation, peritonitis, oesophagomalasia, clinico-pathologic correlation, Boerhaave syndrome

Volume 6 Issue 2 - 2018

Ariyarhatne HTDW,[1] Hulathduwa SR,[1] Bulathsinghela BCARP[2]
[1]Department of Forensic Medicine, University of Sri Jayawardenepure, Srilanka
[2]Colombo South Teaching Hospital, Srilanka

Correspondence: Ariyarhatne HTDW, Senior lecturer, Department of Forensic Medicine, Faculty of Medical Sciences, University of Sri Jayewardenepura, Gangodawila, Nugegoda, Srilanka, Tel 0112758607, Email weerawickramers@hotmail.com

Received: February 16, 2018 | **Published:** April 02, 2018

Introduction

Oesophageal perforation is not a common entity.[1] Underlying causes for perforation are diverse. It could be accidental, iatrogenic and spontaneous or can follow a homicidal act. Most perforations are iatrogenic while spontaneous perforations account only for a 15% minority.[2] Early diagnosis and appropriate treatment are vital for a successful outcome. Failure to diagnose and misdiagnoses are not uncommon as clinical presentation of oesophageal perforation tends to show a wide array of non-specific signs and symptoms which are in common with an acute abdomen, pancreatitis, peptic ulcer disease, myocardial infarction, penumothorax, acute pericarditis, acute aortic dissection and etc.[3] In this case the patient was initially managed with oral antibiotics for acute gastroenteritis as she presented with fever, vomiting and diarrhea as an outpatient and later with an intravenous drip and antibiotics following admission. The oesophageal perforation may be of partial thickness or transmural. The clinical presentation and the management may vary accordingly.

Anatomically, the perforation may be located in the neck, thorax or below the diaphragm within the abdominal cavity. Each anatomical location gives rise to different clinical presentations and management approaches. Oesophagomalasia is an important entity for forensic pathologists as it could lead to misdiagnosis unless it is recognized prudently. It is a postmortem artifact where the lowermost segment of the oesophagus is spontaneously perforated during the postmortem interval mainly due to the effects of gastric enzymes which might leak in to the adjoining lower oesophagus as the sphincter mechanism fails following flaccidity of muscles after death.[4] A postmortem perforation shows no vital reaction[5] and histology is helpful in suspicious cases.[4] A forensic pathologist should always exclude the presence of an artifactual perforation in such presentations.

Case report

The deceased was a 14 year-old mentally subnormal girl with a past history of epilepsy who lived in an orphanage. Despite occasional fits, she was apparently well until three days prior to the death. She developed fever, vomiting and loose stools for which treatment was sought from the out patients' department of a tertiary care hospital. She was admitted to the same hospital by the fourth day of her illness as her condition was further deteriorating despite treatment, where she died within three hours of admission. Medical records indicate prescribing oral antibiotics and an intra-venous infusion of saline only. Intubation had not been performed. An inquest followed by a judicial autopsy was performed as the cause of death was not known and the deceased was an inmate of an orphanage. The girl was averagely built for her age. Features of trauma, dehydration, neglect or malnutrition were not found on external examination. The abdomen was distended. The internal examination revealed a lower oesophageal (sub-diaphragmatic) transmural, longitudinal perforation measuring 7x2.5 cm in size with necrotic borders and approximately one liter of purulent peritoneal fluid (Figure 1), (Figure 2). The diaphragm was intact. Abdominal and thoracic anatomy was unremarkable. No macroscopic features of lung infection or lung pathology were noticed. Neither macroscopic bowel pathology nor trauma to abdominal or thoracic organs was identified. The liver and kidneys appeared pale. The spleen was moderately enlarged and soft.

Few early adhesions were seen around the stomach and bowel loops. The peritoneum and the surface of bowel loops appeared dusky and their shiny and healthy nature was lost. The cause of death was given as generalized peritonitis following perforation of the lower oesophagus. The underlying cause for the perforation was unascertainable.

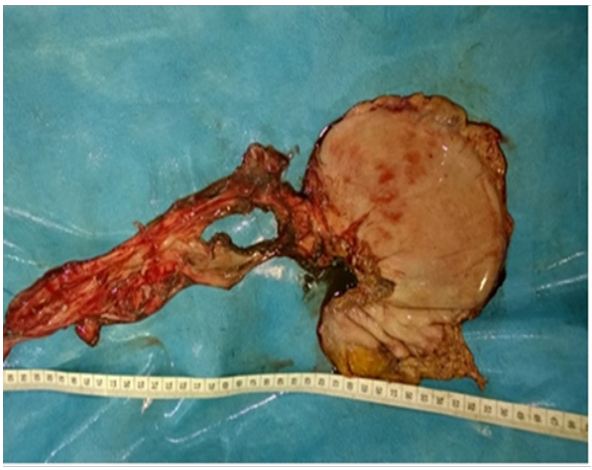

Figure 1 The full thickness perforation of the lower oesophagus with necrotic borders.

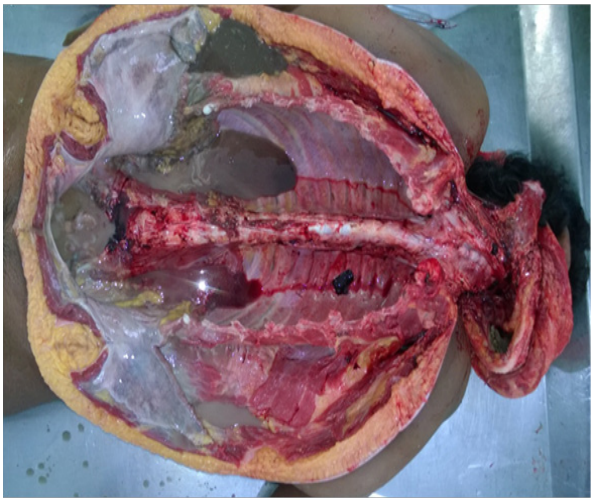

Figure 2 Purulent fluid within the peritoneal cavity.

Discussion

The discussion goes under three main aspects in this case presentation: oesophageal perforation causing peritonitis, the possibility of wrongful identification of oesophagomalasia as an antemortem lesion (and vice versa) and the importance of clinico-pathological discussions regarding the rare post mortem findings revealed during autopsy. The oesophagus could be perforated in cervical, thoracic or abdominal areas due to multiple reasons. The clinical symptoms vary with the corresponding area of damage. The commonest cause for perforation is medical procedures and as such iatrogenic or accidental in origin.[6] The other circumstances that could lead to oesophageal perforation may be due to natural, suicidal and rarely homicidal acts. Except for the medical procedures the other causes that could result in oesophageal perforation are tumors, ulcers caused by gastroesophageal reflux disease (GORD), impaction of swallowed foreign bodies, physical trauma and violent vomiting.[7] In relation with spontaneous rupture of the oesophagus, Boerhaave syndrome should be mentioned: it is due to the rapid rise in the intraluminal pressure after forceful vomiting. It occurs mostly in the lateral, lower 1/3 of the oesophagus.[8] The victims usually present with the symptoms of chest pain, shock or respiratory distress.[9] According to the literature review, Boerhaave syndrome is sometimes termed "effort rupture" of the esophagus: a spontaneous perforation of the oesophagus resulting from a sudden increase in the intra-oesophageal pressure combined with negative intra-thoracic pressure as in the case of severe straining or vomiting.[10] Boerhaave syndrome is associated with the clinical triad of vomiting, chest pain and subcutaneous emphysaema.

This is termed Mackler's triad and is usually seen in 50% of the cases. Other signs and symptoms include epigastric pain, back pain, dyspnoea and shock. It is prudent to suspect a perforated oesophagus in those who present with lower thoracic or abdominal pain in combination with gastrointestinal and respiratory symptoms.[11] The classic presentation of spontaneous rupture includes episodes of severe vomiting or retching followed by severe chest or epigastric pain. Other possible causes would be severe straining, childbirth, weight lifting, bouts of coughing or laughing, intractable hiccups, blunt trauma, seizures and forceful swallowing.[11] This case does not typically fit in to the classical picture of Boerhaave syndrome though there are certain supportive features such as a history of severe vomiting. In typical cases of Boerhaave syndrome there is no peritonitis as the perforation is mostly above the diaphragm. In this case under discussion, the perforation was largely in the sub-diaphragmatic portion of the oesophagus but extension of the perforation towards the sub-diaphragmatic part cannot be excluded in a post postmortem setting. There was no history of trauma, medical interventions or recent fits. Ingestion of any caustic substance or foreign body was not evident. Other risk factors to be considered include a history of preexisting gastrointestinal pathology such as gastro-oesophageal reflux, hiatus hernia, strictures, carcinoma, infection, radiation therapy and Barrett oesophagitis none of which was evident in the history and internal autopsy examination.[3] There was a convincing history that she had episodes of vomiting during the first three days of illness although the severity and frequency of bouts of vomiting were uncertain to be considered as an underlying cause for the perforation.

In addition to the oesophageal perforation, the other major pathology found in this case was generalized peritonitis. Common possible causes for generalized peritonitis such as appendicitis, abscess formation, gynecological causes including possible primary and secondary causes for peritonitis were successfully excluded during the autopsy examination leaving no other pathological condition except for the transmural longitudinal lower oesophageal perforation with necrotic borders measuring 7x2.5 cm in size (Figure 1) as the only plausible culprit. Purulent peritoneal fluid with oesophageal perforation in the absence of other findings provided sufficient grounds to attribute the perforation as the cause for peritonitis (Figure 2). The underlying cause for the perforation was undetermined in this case which is not unusual as most of the causes may be due to a functional aetiology such as building up of a pressure cone during severe straining or vomiting etc. A thorough literature review revealed that cases of spontaneous sub-diaphragmatic rupture of oesophagus are very rarely reported in the world. Such perforations of the abdominal part of the oesophagus present with features of an

Citation: Ariyarhatne HTDW, Hulathduwa SR, Bulathsinghela BCARP. A rare case of sub-diaphragmatic oesophageal perforation resulting in peritonitis. *Forensic Res Criminol Int J.* 2018;6(2):100–102. DOI: 10.15406/frcij.2018.06.00190

acute abdomen, peritonitis and rarely with gastrointestinal bleeding. Attempting to identify the underlying cause for the antemortem oesophageal perforation is a worthwhile exercise.

In this particular case it was not possible to delineate the exact underlying cause for the perforation other than excluding some of the common causes therefore leaving the underlying cause for the rupture as undetermined. The available history suggested that there had been bouts of vomiting during her acute illness to cause sudden increase in intra-thoracic pressure making the possibility of oesophageal perforation as in Boerhaave syndrome but with an atypical presentation. Gastromalasia and oesophagomalasia are two conditions that warrant accurate interpretation by a forensic pathologist differentiating from ante-mortem perforations. Though gastromalasia is a well-documented autopsy artifact, oesophagomalasia has been very rarely documented in the medical literature. Oesophagomalasia appears to the naked eye with slimy brown, thinned out margins devoid of vital reaction. Histology plays a key role in differentiating ante-mortem ruptures from post-mortem artifacts such as gastromalasia or oesophagomalasia both of which fail to show an acute inflammatory response. The usual practice of stating the cause of death alone without any other relevant details on the bed head ticket by the forensic pathologist who performs the autopsy at least in rare conditions such as this case would be grossly inadequate to trigger a clinico-pathological dialogue among the treating clinicians. The rare pathological findings that are only surfaced by the post mortem examinations are worthwhile to be revisited by the clinicians, desirably by making their presence at the autopsy room or by communicating with the forensic pathologist verbally. Such clinico-pathological discussions would prove to be a healthy exercise mutually rewarding for both the clinician as well as the pathologist.

Conclusion

Upon exclusion of traumatic and pathological conditions leading to oesophageal rupture, this case could be concluded as a spontaneous rupture of sub-diaphragmatic oesophagus where the exact cause for the spontaneous rupture was unable to be ascertained. Oesophagomalasia is an entity vital to be excluded by the forensic pathologist before arriving at this conclusion. Findings that are solely unearthed at the autopsy warrant a regular clinico-pathologic correlation by the clinicians and the forensic pathologists as unsuspected and rare entities would be overlooked otherwise.

Acknowledgment

None.

Conflict of interest

None.

References

1. Vidarsdottir H, Blondal S, Alfredsson H, et al. Oesophageal Perforations in Iceland: a Whole Population Study on Incidence, Aetiology and Surgical Outcome. *Thorac Cardiovasc Surg*. 2010;58(8):476–480.
2. Brinster CJ, Singhal S, Lee L, et al. Evolving options in the management of esophageal perforation. *Ann Thorac Surg*. 2004;77(4):1475–83.
3. Esophageal Rupture and Tears in Emergency Medicine Differential Diagnoses. 2016.
4. Aggrawal A. APC Essentials of Forensic Medicine and Toxicology. 1st ed. New Delhi: Avichal Publishing Company; 2014.
5. Bardale R. Principles of Forensic Medicine & Toxicology. New Delhi: JP Medical Ltd; 2011.
6. Commonest cause medical procedure.
7. Esophageal Perforation. Healthline. 2017.
8. de Schipper JP1, Pull ter Gunne AF, Oostvogel HJ, et al. Spontaneous rupture of the oesophagus: Boerhaave's syndrome in 2008. Literature review and treatment algorithm. *Dig Surg*. 2009;26(1):1–6.
9. Aga Z, Avelino J, Darling GE, et al. An Unusual Case of Spontaneous Esophageal Rupture after Swallowing a Boneless Chicken Nugget. *Case Rep Emerg Med*. 2016:5971656.
10. Boerhaave syndrome: Effort rupture of the esophagus.
11. Tonolini M, Bianco R. Spontaneous esophageal perforation (Boerhaave syndrome): Diagnosis with CT-esophagography. *J Emerg Trauma Shock*. 2013;6(1):58–60.