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Esophageal Causes of Sudden and Unexpected Death

ABSTRACT: Gastrointestinal conditions are uncommon causes of sudden and/or unexpected death as compared to cardiovascular diseases, motor vehicle trauma, or suicide, and may involve an array of fatal mechanisms. Lethal esophageal conditions are encountered even less often, the manifestations of which include acute upper airway occlusion from tumors or foreign material, intraluminal hemorrhage from vascular abnormalities, or perforation with fistula formation resulting in hemorrhage and sepsis. When encountered at autopsy, a particular condition may also be a manifestation of a disease that does not primarily involve the esophagus. For this reason, a detailed autopsy investigation is required for evidence of systemic or remote disease when lesions are found within the esophagus. In this report, possible life-threatening esophageal conditions are reviewed with a description of lethal mechanisms, mention of rare associated diseases, and comment on difficulties that may arise at autopsy in the evaluation of such cases.

KEYWORDS: forensic science, sudden unexpected death, esophagus, tumor, varices, foreign body

The predominant cause of sudden natural death in adults in Western countries is atherosclerotic coronary artery disease. Unnatural deaths in regions not involved in armed conflict are most often because of suicides or motor vehicle accidents. In infants and young children, entities such as sudden infant death syndrome (SIDS), congenital diseases, and infections account for a significant proportion of unexpected deaths (1). Primary gastrointestinal conditions represent, therefore, a relatively uncommon cause of sudden and unexpected death at any age. In the following report, possible conditions involving the esophagus causing sudden and/or unexpected death that may be encountered at autopsy are described, with a summary of possible lethal mechanisms, mention of rare associated diseases, and delineation of some of the difficulties that may be encountered in determining such diagnoses at autopsy.

TABLE 1—*Abnormal esophageal findings at autopsy associated with sudden and/or unexpected death.*

Obstruction with compromise of the adjacent airway:
Foreign bodies
Immediate
Delayed
Tumors
Perforation (± hemorrhage)
Foreign bodies
Ulceration/Barrett esophagus
Hemorrhage
Varices
Internal
External
Other vascular abnormalities
Mallory–Weiss tears
Hematoma
Rupture
Booerhaave syndrome
Iatrogenic
Miscellaneous

Overview

Esophageal lesions or conditions that may cause sudden and unexpected death are not commonly revealed at autopsy. Deaths from esophageal pathology usually result from acute upper airway occlusion from tumors or foreign material, intraluminal hemorrhage from vascular abnormalities, or perforation with fistula formation resulting in lethal hemorrhage and sepsis (Table 1).

Obstruction

Foreign Bodies

Foreign body impaction is one example of an acquired condition of the esophagus that may cause rapid death from a variety of mechanisms (2). Impaction of foreign material within the upper esophagus may result in occlusion of the glottis with rapid asphyxiation. If food or foreign matter lodges further down the esophagus, there may be airway compromise because of external pressure from the intraesophageal mass on the membranous part of the trachea resulting in a critical reduction in the diameter of the tracheal lumen. Material wedging in the esophagus is most often food and may occur in the setting of the so-called café coronary syndrome (3,4). In this syndrome an individual suddenly collapses while eating, sometimes in the middle of a conversation. The underlying problem is impaction of a large unchewed food bolus at any level from the oropharynx to the glottis or upper esophagus (Fig. 1). It has also been reported as a cause of death in animals (5).

While the mechanism of death is thought to be asphyxial, the speed of the terminal episode may suggest that reflex vagal cardiac inhibition may also be involved. Predisposing conditions include dementia, poor dentition, other neurological conditions such as parkinsonism, or bulbar or cerebral palsy where there is difficulty coordinating swallowing reflexes, or drug and alcohol intoxication (4,6). Autopsy investigations therefore require both toxicological evaluation of blood, fluids, and tissues and neuropathological assessment. In young children, food impaction may

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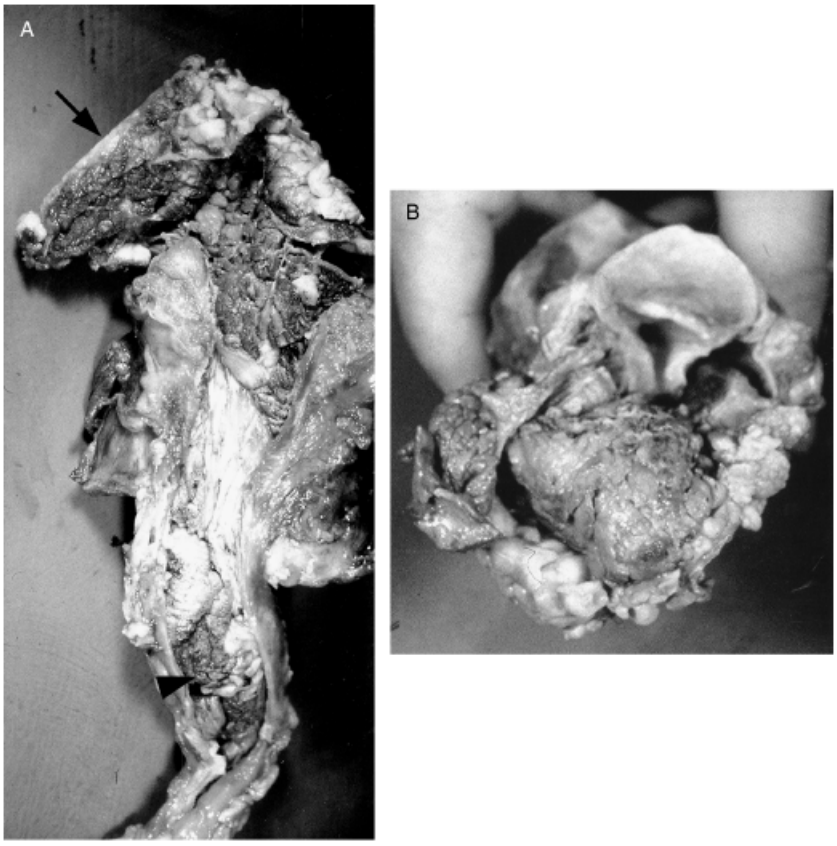

FIG. 1—*Opened esophagus of a 30-year-old male who collapsed at a restaurant while eating, showing two large fragments of poorly masticated meat wedged within the esophageal lumen (arrow—upper esophagus; arrowhead—mid esophagus). (A) A view of the same case showing compression of the glottis from the fragment of meat wedged in the upper esophagus. (B) There was a history of excessive alcohol intake.*

occur because of immature dentition impeding adequate mastication associated with failure to appreciate the size and consistency of an ingested food bolus (7). Particular foods that are associated with an increased risk of choking in young children include those with a firm to hard consistency and a rounded shape such as hot dogs, candy, nuts and grapes (7). Other risk factors in children are conditions such as Treacher Collins syndrome where there is midfacial hypoplasia, cleft palate and malocclusion of teeth, and mental retardation (4). Small rounded toy parts are also associated with upper airway obstruction at young ages.

Although most individuals with significant obstruction manifest immediate symptoms, cases have been described, particularly in infants, where objects such as coins have lodged in the esophagus asymptomatically that have only led to critical narrowing of the adjacent trachea after some time because of compression and inflammation (8). Button batteries may be particularly damaging if they impact in the esophagus because of leakage of corrosives.

Unfortunately, there may be no evidence of the causative obstructing material at autopsy because of removal during attempted resuscitation, or dislodging of the material due to agonal vomiting or swallowing, or during movement of the body. For this reason, it is important to obtain an accurate history from individuals who were at the scene, in addition to ambulance and medical staff who were involved in resuscitative efforts.

Tumors

Critical narrowing of the airway may also be due to intrinsic lesions such as lipomas and fibrovascular polyps. Intraesophageal lipomas are uncommon tumors that may very rarely cause sufficient tracheal compression to compromise respiration (9). Fibrovascular polyps, although benign, are pedunculated tumorlike masses that may reach sufficient size to cause sudden death from laryngeal and glottic occlusion (10). These tumors may also be responsible for hemorrhage. Even when multiple, benign tumors rarely cause acute problems (Fig. 2). If potentially obstructive lesions of the upper aerodigestive tract are encountered at autopsy, the possibility of contributions to luminal narrowing by mucoid secretions because of a coincidental upper respiratory tract infection should be considered. In addition, toxicological screening of blood and fluids may provide important information if there has been narcotic or sedative ingestion that may have contributed to respiratory depression and airway narrowing by causing muscle relaxation and reducing clearance of airway secretions (11).

Perforation

Impacted foreign bodies may also erode through the wall of the esophagus resulting in sepsis or lethal hemorrhage if the adjacent aorta or one of its major branches is perforated (12,13). Victims are often alcoholics or are elderly, demented individuals and wear dentures. Animal bones, such as fish or chicken bones, are likely contenders causing aortoesophageal or carotidesophageal fistulas, the first manifestation of which may be a small "signal" hemorrhage followed by a massive hematemesis and then cardiovascular collapse and death (14). Perforation usually occurs at sites of anatomical narrowing. Symptoms of animal bone perforation of

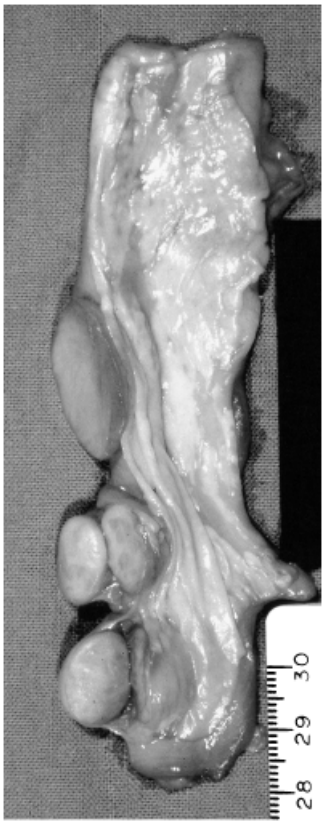

FIG. 2—*Incidental multiple leiomyomas of the esophageal wall.*

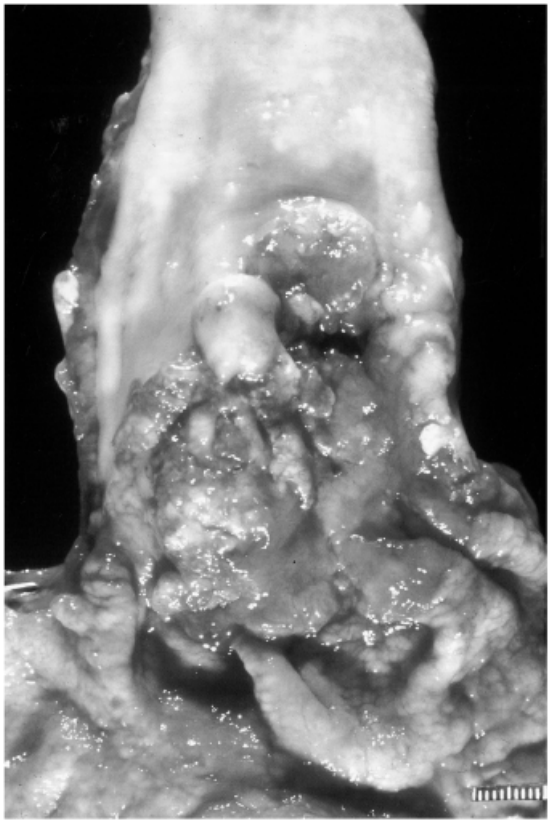

FIG. 4—*Ulcerating and infiltrating carcinoma of the lower esophagus.*

the esophagus, and at other levels of the intestine, may be delayed for months before the development of fatal bleeding or sepsis (15,16). Other complications may include pyopneumothoraces.

Perforation of the esophageal wall may also result from ulceration because of underlying disease. This is most often caused by ulceration that may be associated with Barrett esophagus due to the reflux of acid stomach contents into the lower esophagus. Barrett esophagus is characterized by the replacement of squamous with columnar epithelium in the lower esophagus (Fig. 3) and has an increased risk of adenocarcinoma (17,18). Perforation of the wall of the esophagus has also been associated with penetration into adjacent structures including the airways and pleural spaces (19). Life-threatening esophagobronchial fistula, tension pyopneumothorax, esophagoatrial and esophagoaortic fistulas and massive hemorrhage have been reported. Rarely Barrett esophagus may be associated with other conditions such as CREST (calcinosis, Raynaud phenomenon, esophagitis, sclerodactyly, and telangiectasia) that need to be checked for at autopsy (20–23). Erosion into adjacent vessels may also be associated with carcinoma of the esophagus (Fig. 4) (12), and may complicate surgical repair if there is anastomotic dehiscence. Other causes of esophageal rupture include blunt trauma and rupture of an esophageal diverticulum (21).

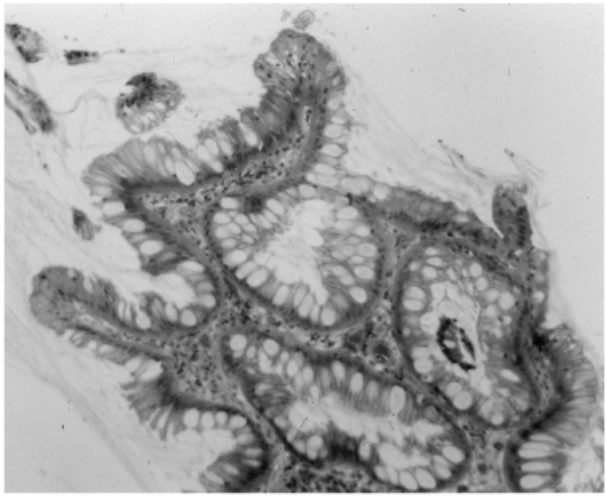

FIG. 3—*Columnar epithelium in the lower esophagus in a case of Barrett esophagus.*

Hemorrhage

Varices

Acute hemorrhage may result from fistulas between the esophagus and major vessels as described above, but is more commonly due to bleeding from a ruptured intraesophageal varix. Varices most often arise from portal hypertension due to liver cirrhosis from a number of entities including alcoholic and primary biliary cirrhosis. Increased venous pressure within the communicating and gastroepiploic veins of the stomach and short gastric veins of the spleen results in varices within the lower esophagus (24). Victims of this condition often present to autopsy with histories of being found dead alone at their home address surrounded by copious amounts of vomited blood and fresh melena. There may also be a history of alcohol abuse with the stigmata of liver failure consisting of jaundice, ascites, peripheral edema, multiple cutaneous bruises, and effusions. There may be

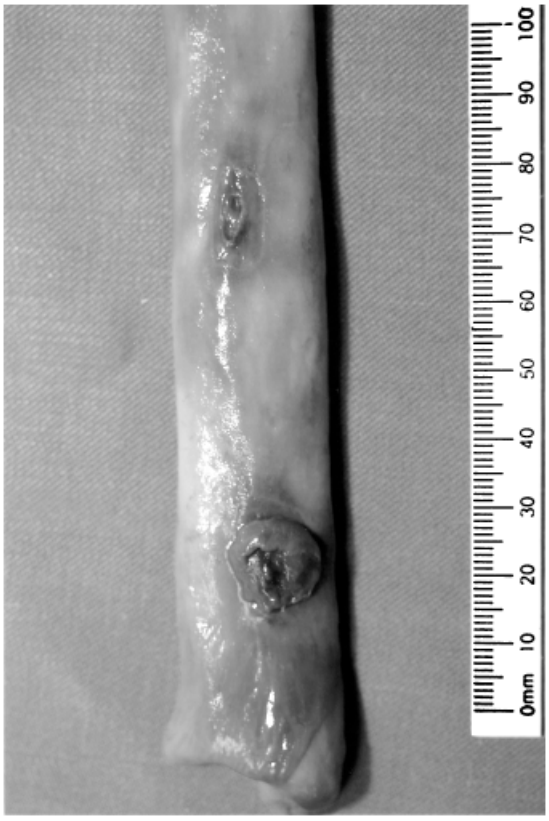

FIG. 5—*Two sites of rupture with bleeding from esophageal varices due to portal hypertension from alcoholic liver cirrhosis in a 47-year-old woman. Adherent blood clot was present around the lower varix.*

blood staining around the mouth with fresh blood within the stomach and upper small intestine. Coagulopathy associated with liver disease may exacerbate the tendency to hemorrhage and toxicology may reveal high levels of alcohol (25,26).

Unfortunately, drainage of blood from varices due to rupture and dissection, combined with postmortem mucosal discoloration, may make identification difficult. Special dissection of the esophagus with tying off and inverting of the esophageal tube will assist in demonstrating varices (27) that can usually be clearly seen histologically. On occasion, there may be more than one bleeding point (Fig. 5). While hemorrhage usually occurs into the esophagus, in rare cases there may be rupture of externally located varices, with death resulting from hemorrhage into one of the pleural cavities (28).

Other Vascular Abnormalities

Although esophageal varices are usually associated with portal hypertension due to liver cirrhosis, this may not always be the case, and varices have been reported due to portal vein thrombosis in the absence of cirrhosis. Portal vein thrombosis may be caused by a variety of other conditions, including sepsis, pancreatitis, metastatic carcinoma, biliary tract surgery, myeloproliferative and thrombotic disorders. In 50% of the cases, it is idiopathic and is usually associated with a benign clinical outcome, although unexpected death may occur (24). Bleeding esophageal varices have also been described as a rare complication of portal hypertension because of sarcoidosis (29). In addition, acute hemorrhage has been reported in a case of gastroesophageal angiodysplasia (30). Thus, the presence of acute hemorrhage from the lower esophagus may not always be due to varices, and if present, varices may not always be associated with liver cirrhosis. This may necessitate careful searching for less common underlying diseases at autopsy.

Mallory–Weiss Tears

Significant acute upper gastrointestinal hemorrhage may also be caused by longitudinal lacerations at the gastroesophageal junction, the so-called Mallory–Weiss tears that were first described by Mallory and Weiss in 1929 (31). While these mucosal tears typically follow an episode of repeated vomiting after an alcohol binge (so-called emetogenic injury), there are a variety of other precipitating events that may cause a sudden increase in intragastric pressure, or intussusceptions of the stomach into the lower esophagus, with resultant mucosal injury. These include vomiting from other causes such as pregnancy, raised intracranial pressure, ipecac, and cytotoxic drug administration, in addition to forceful coughing, straining at stool, snoring, hiccupping, cardiopulmonary resuscitation, and blunt abdominal trauma (32).

Intramural Hematoma

Vomiting may also be associated with intramural hematoma formation within the esophagus (esophageal apoplexy) that may lead to massive hemorrhage (33). Intramural hematoma has an association with underlying coagulation disorders or the use of anticoagulant drugs.

Rupture

Boerhaave Syndrome

A related phenomenon, spontaneous rupture of the esophagus was first described by Boerhaave in 1724 and is associated with a significant mortality rate (Fig. 6). It is also believed to result from a sudden increase in intraluminal pressure during forceful vomiting and is not usually associated with significant hemorrhage (34,35). It commonly follows excess alcohol consumption and so toxicology may be useful at autopsy. Other precipitants include childbirth, weight lifting, convulsions, bulimia, defecation, laughing fits, severe asthma, and prolonged coughing or hiccuping. It may occur in the newborn or follow herpetic esophagitis (34,35). Complications include sepsis and pleural effusions with or without pyopneumothorax (36). Urgent surgery may prevent death, although Boerhaave believed it to be a "deadly malady" and advised: "Let not anyone insanely suggest that the right intercostal regions should be perforated by the Surgeon. Nor let it be said that the humors in the thoracic cavity could be emptied" (34).

Iatrogenic Causes

A variety of iatrogenic events may precipitate in esophageal rupture including esophagoscopy, sclerotherapy, intraesophageal tube placement, and dilatation of esophageal strictures. Acquired strictures of the esophagus usually result from ingestion of caustic substances in childhood with resultant scarring and narrowing of the esophageal luminal diameter. Subsequent treatment often necessitates repeated dilatations. In one study, of the 195 patients requiring esophageal dilatations, six (18%) died, some rapidly because of pneumothorax with mediastinal shift (37).

Miscellaneous

Sudden death from aortoesophageal fistula complicating recent caustic ingestion has been described (38), as has lethal aortic

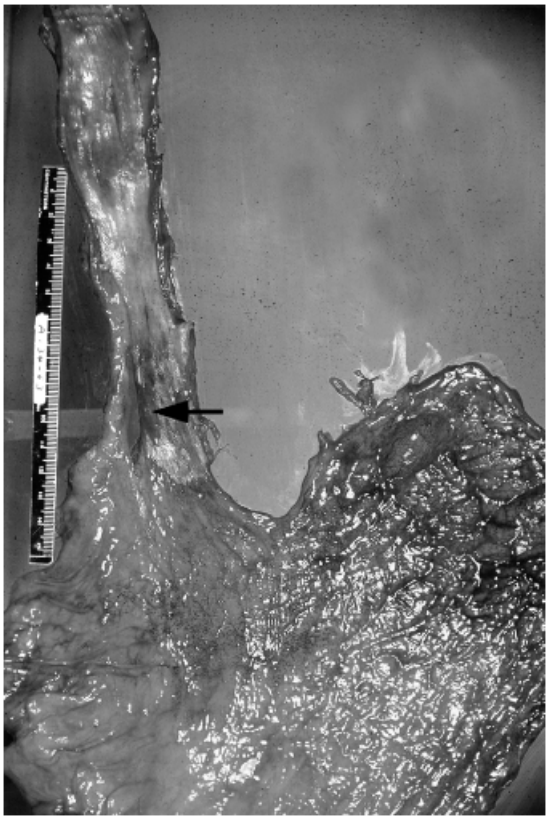

FIG. 6—*Spontaneous rupture of the distal esophagus (arrow).*

rupture in a 42-year-old male following surgery and radiochemotherapy for squamous cell carcinoma of the distal esophagus (39). Other intrinsic diseases of the esophagus have rarely been associated with sudden death including achalasia, a motor disorder of the esophagus where there is increased tone in the lower esophageal sphincter with dilatation of the adjacent esophagus. This predisposes to aspiration and rarely to possible cardiac arrhythmia associated with widening of QRS complexes, high-grade atrioventricular block, and ventricular asystole (40). Heterotopic gastric mucosa has also been reported as predisposing to aspiration (41).

Summary

There are a wide variety of causes of sudden death due to intrinsic and extrinsic lesions of the esophagus that involve airway obstruction, perforation, pneumothorax, hemorrhage, and sepsis. Demonstration of causative conditions or lesions at autopsy may not be straightforward, and esophageal problems, while lethal, may merely be a manifestation of more generalised or distant disease, the features of which need to be looked for at autopsy.

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