Arterial-Esophageal Fistulae Developing in Patients with Anomalies of the Aortic Arch System


Two cases are presented in which anomalies of the aortic arch system were associated with development of an arterial-esophageal fistula. The fistula resulted in massive upper gastrointestinal hemorrhage and death. In each malformation, part of the anomalous aortic arch system lay against the esophagus and thereby provided the anatomic substrate for an arterial-esophageal fistula. In both cases, nonmassive ("sentinal") hemorrhage occurred prior to the massive fatal hemorrhage. Recognition of the significance of the "sentinal" hemorrhage may allow surgical correction of the problem avoiding uncontrolled massive hemorrhage.

The most common causes of hematemesis are gastroduodenal ulcers and esophageal varices. Less common disorders, however, including arterial-esophageal fistulae, may also produce hematemesis. Their rapid clinical recognition may be necessary to prevent massive fatal hemorrhage.

Arterial-esophageal fistula represents an uncommon, often catastrophic complication of either esophageal or arterial diseases. Among the esophageal conditions from which a fistulous tract may develop are esophageal cancer, lye ingestion and penetration by foreign bodies. Among states of the aortic arch or its branches which may be complicated by the development of an arterial-esophageal fistula are thoracic aortic aneurysm, aneurysm of the branches of the aorta, and prostatic aortic grafts.

We observed two exceptional cases in which patients with congenital anomalies of the aortic arch system developed an arterial-esophageal fistula. We are prompted to place these cases on record for several reasons, including recognition of factors which may have potentiated the development of the fistula, the possibility for surgical correction, and the unusual nature of the cases.

Case Reports

Clinical Features

A 36-year-old black woman presented to her local physician with history and physical findings of acute subarachnoid hemorrhage.

A lumbar puncture was performed and yielded xanthochromatic cerebral spinal fluid. Four-vessel cerebral angiogram showed a berry aneurysm of the posterior communicating artery. Right aortic arch with aberrant left subclavian artery was also demonstrated. The patient was then transferred from a community hospital to United Hospitals, St. Paul, Minnesota, for surgical treatment of the cerebral aneurysm.

On admission to United Hospitals, physical examination revealed an alert, obese woman. Blood pressure was 130/80 mm Hg and was equal in both arms. Neurologic examination revealed ptosis and mydriasis of the left eye and slightly hyperactive left Achilles tendon reflex.

After treatment with dexamethasone, aminocaproic acid and phenobarbital for six days, the patient was operated upon and the aneurysm was clipped.

Postoperatively, the patient's condition was unstable and complicated by cardiac arrest. Following cardiopulmonary resuscitation, the patient never regained consciousness. CT scan of the brain disclosed a left frontal hematoma. Because of deterioration in respiratory function, the patient received mechanical ventilation. A nasogastric tube was placed through which feeding was accomplished. Dexamethasone was administered during the patient's entire hospital course, initially orally and then parenterally. The dose ranged between 4 mg every 6 hours and 10 mg every 8 hours. No anticoagulant was administered.

Seventeen days following the initial operation upon the cerebral aneurysm, the patient was reoperated, and a hematoma in the left frontal lobe was evacuated. Returns from the nasogastric tube were noted to contain blood.

Twenty-seven days following the initial neurosurgical operation, massive hematemesis developed. Emergency endoscopy was performed, and active hemorrhage in the esophagus was noted. The stomach and duodenum were normal. The bleeding was uncontrollable. The patient exsanguinated and died.

Pathologic Features

In addition to anomalies of the aorta to be described, significant
findings at autopsy included subarachnoid hemorrhage, an evacuated hematoma in the left frontal lobe, and multiple small bilateral pulmonary emboli.

The intracardiac structures were within normal limits. A right aortic arch with retroesophageal segment and aberrant left subclavian artery were present. The branches of the aortic arch (from proximal to distal) were left common carotid, right common carotid, right subclavian and left subclavian arteries. The left subclavian artery arose from a diverticulum of the aorta (remnant of the left aortic arch) at the junction of the right aortic arch and descending thoracic aorta. The aortic diverticulum lay against the left-posterior aspect of the esophagus. A left ligamentum arteriosum was present and ran between the left pulmonary artery and the aforementioned aortic diverticulum, forming a vascular ring.

A fistulous tract was present between the esophagus and the aortic diverticulum (Fig 1). Histologic examination of the aortic diverticulum at the level of the fistula demonstrated necrosis of the arterial wall with infiltration of leukocytes, mainly neutrophils, and to a lesser extent lymphocytes and plasma cells. At the same level, the esophagus showed transmural necrosis with a cellular infiltrate similar to that in the wall of the aortic diverticulum (Fig 2). Examination of the fistulous tract with special stains showed Gram-positive cocci and Candida.

**CASE 2**

**Clinical Features**

A 74-year-old man was noted by thoracic roentgenogram to have a mass in the right upper hemithorax interpreted as a tortuous innominate artery. The patient was asymptomatic, and the mass, followed radiographically, was noted over the course of several years to increase slightly in size (Fig 3). At the age of 79 years, the patient complained of dysphagia. Barium esophagram disclosed a mass projecting against the mid-portion of the posterior aspect of the esophagus. Esophagoscopy disclosed extraluminal posterior compression of the esophagus. CT scan of the thorax was interpreted as showing a left aortic arch with a large aneurysm at the origin of an aberrant right subclavian artery. Surgical treatment of the aneurysm was offered, but the patient refused.

At 81 years of age, the patient sought medical consultation because of black, tarry stools. Before evaluation could be initiated, massive hematemesis occurred. Emergency intubation was accomplished, and a nasogastric tube was inserted. Emergency surgical exploration of the thorax disclosed the aberrant right subclavian artery. The origin of the artery was involved by an aneurysm that lay adjacent to the posterior wall of the esophagus. In an attempt to dissect the vessel free from adjacent structures, the cavity of the aneurysm was entered. It contained foul-smelling, purulent debris and a coiled portion of the previously placed nasogastric tube. The patient exsanguinated and died during the operation.

**Pathologic Features**

At autopsy, significant findings were related to the aortic arch

**FIGURE 1.** Case 1. View of thoracic organs from behind. The esophagus has been opened. The right aortic arch has passed over the right main bronchus to join the left-sided descending aorta. Arising from the aorta at the junction of the right arch and descending portion is a diverticulum from which the left subclavian artery arises. The aorta has been separated somewhat from the esophagus in dissection. The black probe shows the site of communication between the aortic diverticulum and the esophageal lumen. D = diverticulum; Des A = descending aorta. E = esophagus. LL and RL = posterior aspects of left and right lungs, respectively. LS = left subclavian artery; RAA = right aortic arch.

**FIGURE 2.** Case 1. Low power photomicrograph of aortic diverticulum at the site of the fistula. The general position of the fistula between the diverticulum and the esophagus lies between the two pairs of arrows. In this tract, exudate is present associated with transmural necrosis of the atrial wall (H & E, original magnification X 5).

**FIGURE 3.** Case 2. Posteroanterior thoracic roentgenogram taken approximately two years before death. The large mass is caused by the aneurysm of the aberrant right subclavian artery.
Arterial-Esophageal Fistulae (Edwards et al)

In the series of Carter et al of 24 patients with aortoesophageal fistula, 80 percent exhibited a minor premonitory upper gastrointestinal hemorrhage termed the "sentinal" hemorrhage. According to Carter and others, there exists a period of time ("grace period") between the "sentinal" hemorrhage and the occurrence of massive exsanguinating hemorrhage. During this time, the fistula may be diagnosed and amenable to surgical repair.

In the study of Carter et al of 24 cases of aortoesophageal fistula, there was no case associated with anomalies of the aortic system. The backgrounds for the fistulae were thoracic aortic aneurysms in 16 cases and thoracic neoplasms (either esophageal or bronchogenic) in four cases. In the remaining four cases, the causes varied and included esophageal foreign body. Cerestin feeding tube perforation, benign esophageal ulcer and atheromatous aortic ulcer in one case each.

While the etiology of the fistula in Case 1 may never be proved, it is possible to speculate as to its pathogenesis. The vascular ring in this case may have applied transmural pressure to the adjacent esophagus, and the nasogastric tube may have caused trauma and ulceration. If such ulceration occurred in relation to the adjacent artery, there would be the potential for a fistula to develop between the artery and the esophagus. Additionally, the negative influence which corticosteroids have on inflammatory responses may have allowed tissue injury and secondary infection without the usual response. In such a setting the potential for the development of a fistulous tract may have been enhanced.

In Case 2, an arterial-esophageal fistula developed from an aneurysmal segment of an aberrant right subclavian artery. We do not know of any potentiating factors for fistulous development in this case aside from the pulsating aneurysm. Aneurysmal erosion into the gastrointestinal tract is not unusual. This phenomenon is most often observed in cases of abdominal aortic aneurysm which communicate with the overlying duodenum. What is unusual about Case 2 is the location of the arterial-enteric fistula. The retroesophageal course which the aberrant subclavian artery followed provided the anatomic substrate for such a fistula.

In this report, two cases are presented in which an arterial-esophageal fistula developed in patients with anomalies of the aortic arch system who had been asymptomatic most of their lives. Recognition of "sentinal" hemorrhage and prompt surgical intervention could have potentially influenced the course in these patients.

**Comment**

In the two cases presented, congenital anomalies of the aortic arch system allowed direct contact between the arterial system and the esophagus. Under these circumstances, an arterial-esophageal fistula developed and resulted in massive hemorrhage into the esophagus.

In Case 1, a diverticulum of the aorta lay against the esophagus as it arose from the junction of a retroesophageal segment of a right aortic arch and the descending aorta. In Case 2, there was an aneurysmal segment of an aberrant right subclavian artery lying against the posterior aspect of the esophagus, later communicating with the esophageal lumen.

It is of interest that in each case prior to the massive hemorrhage the patient had evidence of nonmassive upper gastrointestinal hemorrhage. We believe that this represents the phenomenon of the "sentinal" hemorrhage first described by Chiari in 1914 and stressed by Carter and associates in 1978.

Figure 4, Case 2. Transverse section of the aortic arch and adjacent structures viewed from below to simulate a tomographic projection. The right subclavian artery arises as the fourth branch of the aortic arch. The vessel passes posterior to the esophagus, where it becomes aneurysmal. The fistulous tract between the aneurysm and the esophagus is indicated by the white arrow. An = aneurysm; Ant = anterior; Ao Arch = aortic arch; E = esophagus; Post = posterior; RS = right subclavian artery; T = trachea.

System. The aortic arch was left-sided. The branches of the aortic arch from before backward were the right common carotid, the left common carotid, and left subclavian and the right subclavian arteries. The right subclavian artery arose from the right aspect of the junction between the aortic arch and the descending aorta. As the vessel coursed to the right, it passed posteriorly to the esophagus. The proximal portion of the aberrant right subclavian artery was involved by a large aneurysm measuring $8 \times 6 \times 6$ cm. The aneurysm had eroded into the esophageal lumen resulting in a fistula measuring about 1 cm in diameter (Fig 4). Microscopically, there was no evidence of arteritis. The entire aorta demonstrated grade 4 atherosclerosis. With the exception of the esophageal lesion described, the entire gastrointestinal tract was normal, and no other source for melanotic stools was found.
REFERENCES
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6 Edwards JE. Malformations of the aortic arch system manifested as "vascular rings." Lab Invest 1953; 2:56-75

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