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Clinico-Pathological Correlation

Fatal aortoesophageal fistula in two cases of tight vascular ring

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Abstract Vascular rings are rare vascular congenital anomalies causing oesophageal and tracheal compression. An aortoesophageal fistula is a devastating, in part iatrogenic, complication of vascular rings. It is seen with increasing frequency, and can be misleading, since differential diagnosis with other causes of haematemesis and melaena is often difficult, especially in infants. We report two infants with aortoesophageal fistulas secondary to double aortic arches forming a vascular ring. In both, the diagnosis was missed, and massive haemorrhage led to death. In both cases, the fissuration on the oesophageal and aortic sides of the fistula had sharp edges, highly suggestive of an iatrogenic laceration caused by manipulation of nasogastric tubes. The key for the diagnosis of vascular rings is, therefore, clinical suspicion and awareness of this condition. Prompt identification in infants with stridor, wheezing, or respiratory distress can prevent prolonged intubation, thus avoiding the formation of an aortoesophageal fistula and hopefully preventing a fatal outcome.

Keywords: Vascular ring; aortoesophageal fistula; double aortic arch

VASCULAR RINGS ARE RARE VASCULAR congenital anomalies causing oesophageal and tracheal compression. They are produced by a variety of malformations, of which double aortic arch is the most common. The rings usually become clinically manifest during the first months of life. The incidence of symptomatic vascular rings is low and, despite typical symptoms, diagnosis is very often missed, delaying life-saving surgical correction.

An aortoesophageal fistula is a devastating, in part iatrogenic, complication of vascular rings. It is seen with increasing frequency and can be misleading, since differential diagnosis from other causes of haematemesis and melaena is often difficult, especially in infants. It seems to have a progressive course, and can be aggravated by repeated manipulations, such as endotracheal intubation and insertion of nasogastric tubes or endoscopes. Increased awareness is necessary for improving the diagnosis and avoiding delays in treatment. In this report, we describe our experience with two recent cases, both with fatal outcomes.

Clinical histories

Our first case was a 3-month-old girl with a history of growth impairment who presented with episodes of wheezing and stridor attributed to bronchial spasm. The diagnosis of a perimembranous ventricular septal defect had been made at birth. She underwent surgical closure at two-and-a-half months of age. The presence of a right aortic arch had also been detected at preoperative cardiac catheterization. The postoperative course was characterized by haemodynamic instability, with hypotension, bradycardia, and arterial desaturation, requiring intravenous infusion of inotropic drugs for two days. Weaning from mechanical ventilation was impossible, and there were recurrent episodes of bronchial spasm. Cardiac catheterization 1 week after the operation revealed persistence of the left superior caval vein

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and unroofing of the coronary sinus, with a wide interatrial communication through the mouth of the sinus. A residual interventricular communication, along with indirect signs of pulmonary hypertension, was also seen on echocardiography. The patient was kept on mechanical ventilation because of persistent respiratory distress and wheezing. On the 23rd postoperative day, 30 min after replacement of the nasogastric tube, there was profuse and unstoppable haemorrhage from the mouth and nose that terminated with death.

Our second case was a 3-month-old boy. Immediately after a non-complicated birth at term, he presented with respiratory distress, was mechanically ventilated, and was brought to the intensive care unit. A small ventricular septal defect was seen on echocardiography. Mild protrusion of the tracheal mucosa into the lumen, 1 cm over the carena, was seen on bronchoscopy. He was treated with corticosteroids, and was discharged two weeks later. After a few days, the patient was readmitted because of deterioration in his general status. Bacterial endocarditis due to Staphylococcus aureus, with sepsis, meningitis and cerebral infarction secondary to embolization, was diagnosed and treated with antimicrobial therapy. He underwent endotracheal intubation followed by tracheostomy. A nasogastric tube was also inserted, and kept in place during the entire stay in hospital. The patient underwent repeated blood transfusions for persistent anaemia attributed to sepsis. Nearly 2 months later, he presented haematemesis and melaena. He was treated with irrigation of cold epinephrine-saline solution, omeprazol, somatostatin, and blood transfusions. Haematemesis ceased for four days, but after that active bleeding recurred. Upper gastrointestinal endoscopy initially showed massive blood clots in the stomach. The source of bleeding was not clearly identified, but was thought to be a deep gastric ulcer hidden beneath the clots, secondary to cortisone therapy or sepsis. Three days later, two linear gastric ulcers were seen on endoscopy. No oesophageal bleeding was ever seen, since at removal of the nasogastric tube in order to carry out endoscopy, bleeding was so massive as to interfere with evaluation of the oesophageal mucosa. The patient underwent surgical treatment of the ulcers, but developed disseminated intravascular coagulation, and died.

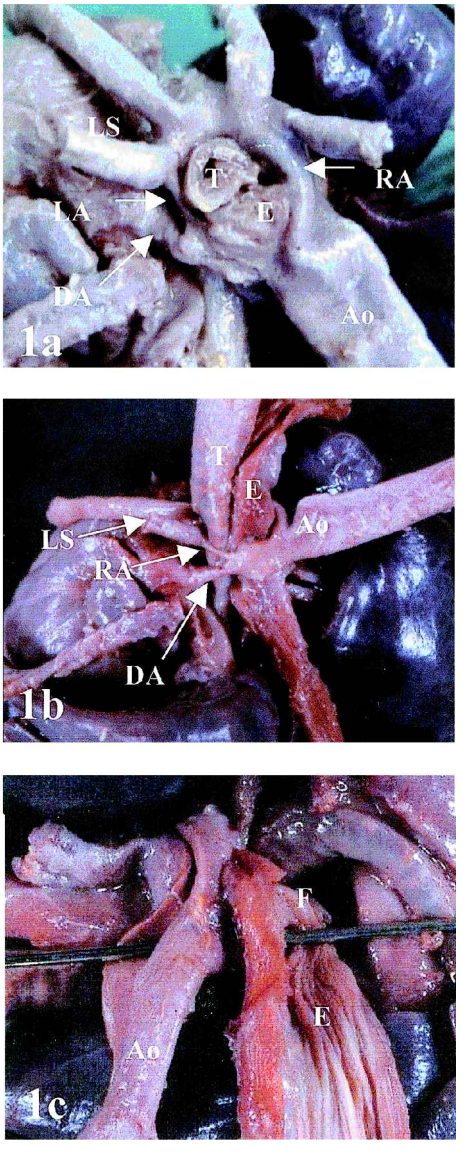

Figure 1.
The autopsied specimen from our first patient. The vascular ring surrounding the oesophagus (E) and trachea (T) is seen from above (a) and the left (b). The thin, atretic segment of the left aortic arch (LA) is evident between the left subclavian artery (LS) and the descending aorta (Ao). Tight adhesion of the oesophagus to the aortic arch is evident. Communication between aorta and oesophagus is demonstrated in (c). DA = arterial duct.

Pathologic findings

The autopsy of the first patient (Fig. 1) revealed persistence of the left superior caval vein draining into the unroofed coronary sinus. A patch had been placed to close the perimembranous ventricular septal defect, leaving a small residual subaortic communication. A vascular ring was found, composed of a double aortic arch, with right carotid and subclavian arteries arising from the right arch, and left carotid and subclavian arteries with separate origin from the left arch. The left aortic arch was atretic distal to the subclavian artery, appearing as a thin fibrous chord connected to the descending aorta by a diverticulum to which the patent arterial duct was also attached. The posterolateral wall of the oesophagus was adhered to the aortic diverticulum by means of fibrous tissue and, between the two,

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there was a fistula 2–3 mm in diameter. The trachea, bronchuses and lungs were filled with blood and mucus.

The autopsy of the second patient (Fig. 2) revealed massive gastrointestinal bleeding due to an aorto-esophageal fistula. The fistula was secondary to a vascular ring consisting of a double aortic arch, with both arches of approximately the same size. Tight constriction of the oesophagus and trachea was present. The oesophagus was adherent to the left aortic arch through abundant fibrous tissue. The arterial duct, still patent, was inserted into the left aortic arch distal to the take-off of the left subclavian artery. The trachea presented severe narrowing 1 cm proximal to the carina, with malacia of the walls. Vegetative endocarditis of the tricuspid and mitral valves was seen, the mitral valve also showing mild dysplasia. Multiple splenic, hepatic and renal infarctions were present, as well as cerebral abscesses in the right temporal lobe.

In both cases, the fissuration on the oesophageal and aortic sides of the fistula had sharp edges, highly suggestive of an iatrogenic laceration caused by manipulation of the nasogastric tubes.

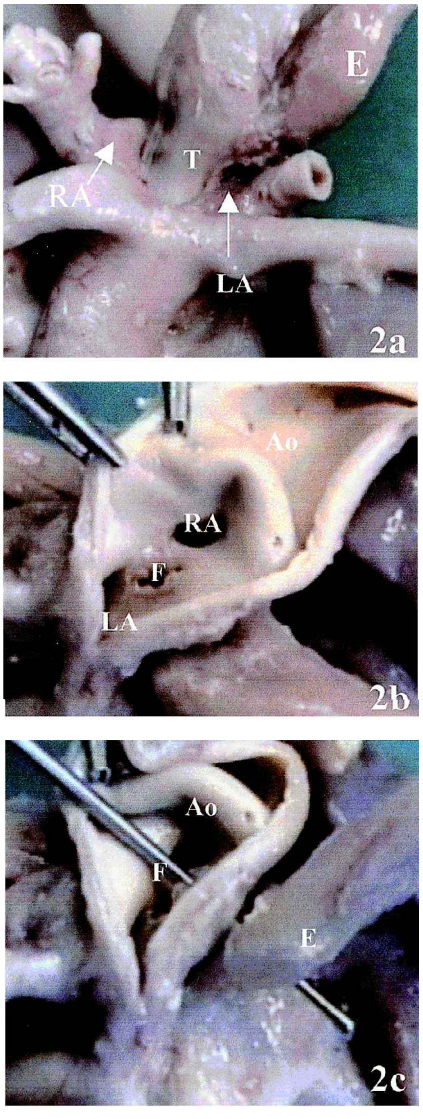

Figure 2.
The autopsied specimen from our second patient. (a) The right (RA) and left (LA) aortic arch are seen – note the tracheal stenosis and tracheomalacia. (b) The fistula (F) from the aortic lumen, situated between the orifice of the right (RA) and left (LA) aortic arches. (c) The direct communication between the aorta and the oesophagus. DA = arterial duct, Ao = descending aorta.

Discussion

The incidence of vascular rings in the general population is currently unknown. Bronshtein et al.[1] used transvaginal ultrasound for the identification of fetal vascular rings in early pregnancy, identifying 6 cases in a population of 5896 fetuses, and giving an incidence of about one case per thousand. Review of the cases corrected in large medical centers confirms the low incidence of vascular rings referred for correction.[2–4] One registry of autopsied cases of congenital cardiac disease reported 21 cases out of a total of more than 6300,[5] whereas in our registry there were 5 cases out of 1360, including the ones reported.[6]

Clinical diagnosis of vascular rings is difficult, even though most patients present typical symptoms. Respiratory distress, apnoea, wheezing, or stridor have been described in most cases.[7,8] Complications of vascular rings are tracheomalacia and frequent lower respiratory tract infections. To the best of our knowledge, 13 cases of aortoesophageal fistula have been described in the English literature.[8–15] Double aortic arches were present in 8 of these cases, of which only 5 survived.

As quoted by Heckstall et al.,[19] Chiari described the clinical presentation of an aortoesophageal fistula due to ingestion of a foreign body as the triad of mild thoracic pain, sentinel arterial haemorrhage, and fatal exsanguination after a short, symptom-free period lasting hours to days. This triad also seems to be common to most patients having aortoesophageal fistula secondary to congenital abnormalities, in whom sentinel bleeding can become manifest as mild haematemesis with bright red arterial blood or as melaena.

Even though fetal echocardiography for prenatal diagnosis of congenital heart disease is today widely used, its utility in the diagnosis of vascular rings has not yet been established. The aortic arch is often difficult to visualize, especially in cases with pleural

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effusion, when there is displacement of organs, or in the setting of a right-sided aortic arch.[16] Transvaginal ultrasound is certainly not suitable for wide-scale screening or for the gathering of epidemiologic data.[1] After birth, barium oesophagography is the traditional means for diagnosis of vascular rings, and some surgeons still greatly rely on this methodology. Computerised tomographic scanning, and especially magnetic resonance imaging, now provide details on the structure of the rings, but can be time-consuming, especially in cases of aortoesophageal fistula.[3] Echocardiography is useful but, like angiography, it often fails to identify atretic segments and is greatly operator-dependent.[4,7,8,13,18] Bronchoscopy is also important in the differential diagnosis of infants with stridor or wheezing. Should there be a vascular ring, then it is possible to visualise the extrinsic compression, often pulsatile, of the trachea by the arterial structures.[4,13]

In our cases, like in most others reported in the literature, the presence of a vascular ring was not suspected. Nasogastric tubes were introduced and kept in place for long periods of time. Prolonged endotracheal intubation was necessary in both cases, often with difficulty encountered in inserting the tube. Tracheal stenosis was repeatedly seen in our second patient during tracheoscopy, but extrinsic compression was never suspected. Prolonged simultaneous endotracheal and enteric intubation presumably caused compression of these structures against the vascular rings. In both cases, abundant fibrous tissue was present, with adherence of the posterolateral wall of the oesophagus to the aorta. This seems to be the site of major friction between the two structures, and corresponds with the site of oesophageal compression as seen on barium oesophagography. The direct relation between the presence of the nasogastric tube and the formation of the aortoesophageal fistula was clearcut in our first case, in which profuse bleeding developed shortly after replacement of the nasogastric tube. Moreover, the two linear ulcers seen in the stomach of our second patient were probably secondary to manipulation of the nasogastric tube.

Endoscopy is currently the most efficient means for diagnosis of upper gastrointestinal bleeding. Nevertheless, direct visualization of the site of bleeding site is often difficult, especially when bleeding is of arterial origin. In our second patient, removal of the nasogastric tube during endoscopy caused profuse haemorrhage within the oesophagus and stomach, on the one hand making precise localization of the source impossible, and on the other hand revealing a "plugging" effect of the nasogastric tube on the fistula. Endoscopy, nonetheless, can readily exclude other more common causes of gastrointestinal bleeding, focusing our attention on intrathoracic sources.[8]

Sentinel bleeding was seen in our second patient. The mechanism responsible for the lag between the sentinel bleeding and the onset of profuse haemorrhage may be the combined result of local "spasm", thrombosis of the fistula, and a "plugging" effect of the nasogastric tube against the longitudinal plication of the oesophagus. Gastrostomy should be done when long-term gastric nutrition or drainage is necessary.[15]

The key to diagnosis of vascular rings, therefore, is clinical suspicion and awareness of the condition. Prompt identification in infants with stridor, wheezing, or respiratory distress can prevent prolonged intubation, thus avoiding the formation of an aortoesophageal fistula and hopefully avoiding the fatal outcome as witnessed in our cases.

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