Mind the fistula

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Thoracic aortic dissection (TAD) with aorto-oesophageal fistula formation is an uncommon but fatal cause of haematemesis. A case is presented of a previously healthy 63-year-old man who presented to the emergency department with syncope and haematemesis. He had no history of heavy alcohol intake and was previously well. Bedside portable chest x ray revealed a widened mediastinum and, while awaiting a CT angiogram, he had a further large haematemesis which led to him undergoing resuscitation and rapid sequence intubation. CT angiography of his thoracic aorta revealed a large thoracic aortic aneurysm with dissection from the arch of the aorta down to the left iliac artery. The dissection had formed an aorto-oesophageal fistula in the proximal oesophagus. He was transferred to a tertiary referral centre for repair of the dissection and aorto-oesophageal fistula.

A 63-year-old man presented to the emergency department complaining of a sudden onset of feeling unwell, followed by a syncopal episode while seated and an episode of vomiting bright red blood. In addition, he complained of a 15 week history of interscapular back pain which was being managed in the community with Co-codamol 30/500. Despite this, the pain had persisted. He gave no history of alcohol abuse or upper gastrointestinal symptoms. He denied any abdominal pain or any history of melaena. Review of his systems revealed only that he had experienced a recent history of dysphagia to solids. His past medical history was significant for hypertension and hypercholesterolaemia and he was a smoker of one or two cigarettes per day and denied any illicit drug use.

On physical examination, the patient was pale, diaphoretic and clinically shocked. Despite this, his vital signs remained within normal limits. He received 500 ml of 0.9% saline by the ambulance crew as his initial blood pressure on scene was noted to be 96/54 mm Hg. Subsequent blood pressure checks in his left and right arms revealed a blood pressure of 127/87

and 115/74 mm Hg, respectively. The cardiovascular examination revealed no murmurs, rubs or gallops and his lung sounds were clear to auscultation. His abdominal examination revealed a soft, non-tender abdomen. His neurological examination was grossly intact.

A venous blood gas, obtained while the patient was breathing room air, revealed a pH of 7.13, pCO₂ 8.23 kPa, pO₂ 4.37 kPa, HCO₃ 20.1 mmol/l, BE -9.0 mmol/l, haemoglobin 9.4 g/dl and haematocrit 31.7%. An ECG revealed normal sinus rhythm only. His portable chest x ray revealed a widened mediastinum. While awaiting CT aortogram, the patient had a large haematemesis, 800 ml of fresh blood, and became increasingly pale, diaphoretic and agitated. He was resuscitated and underwent rapid sequence intubation with etomidate and suxamethonium and was given type O negative blood and intravenous omeprazole. He was transferred for CT aortic angiography which revealed a large thoracic aortic aneurysm measuring approximately 8 cm in AP diameter. There was a prominent Stanford type B dissection within it which appeared to be arising from the aortic arch and progressing along the descending aorta down to the left iliac artery. There was a tongue of extension of contrast into the central mediastinum near the oesophagus, and leakage into the oesophagus was radiologically suspected (figure 1). The cuff of the endotracheal tube was sitting at the level of the fistula and may have contributed to occluding the fistula.

The patient was resuscitated to a goal systolic blood pressure of 80 mm Hg and was transferred to a tertiary cardiothoracic surgery centre. He underwent left posterolateral thoracotomy, resection of the descending aortic aneurysm and replacement with a 34 mm Hemashield graft. The aorta was aneurysmal from the arch to the diaphragm. The fistula entry point was identified at the level of the carina. There was no obvious spinal artery origin in the false lumen (figure 2).

Figure 1 Chest x ray revealing the widened mediastinum.

Figure 2 Parasagittal CT image of the thorax and abdomen showing the thoracic aortic aneurysm and aortic dissection throughout its length.

Primary aorto-oesophageal fistula with thoracic aortic aneurysm and dissection is a highly lethal entity which is often extremely difficult to diagnose. A literature search of the CINAHL, Embase and MEDLINE databases using the search terms ‘arteriosclerosis’, ‘fistula’, ‘aortic diseases’, ‘aortic aneurysm’, ‘esophageal fistula’, ‘aorta aneurysm’ and ‘aortointestinal fistula’ returned four key references.^{1–4}

The diagnosis of thoracic aortic disease (TAD) is often delayed. Viljanen⁵ examined the incidence of diagnostic delays in TAD and found a delay of more than 24 h in 53% of cases of type B TAD. An appreciation of the

risk factors for TAD may lead to fewer missed cases. Some well described risk factors include hypertension, old age and male gender. However, some recently described risk factors, such as cocaine use, pregnancy and sleep apnoea, need consideration if this important diagnosis is not to be overlooked. Other, rarer, risk factors include the connective tissue diseases Ehlers–Danlos and Marfan’s syndrome, bicuspid aortic valve, as well as weight lifting and recent Ecstasy use.

A good history and physical examination, in particular paying attention to the quality, radiation and intensity of the pain, may be helpful in evaluating a patient for TAD. Klompas⁶ evaluated the accuracy of the history and physical examination and found that a focal neurological deficit associated with ‘ripping’ chest pain and migrating pain had the highest likelihood ratio in predicting TAD. An awareness of the atypical ways in which TAD can present, in particular seemingly disconnected symptoms above and below the diaphragm and painless syncope, are recognised presentations of TAD. In our case, both the history of interscapular pain, dysphagia to solids and the widened mediastinum on chest x ray were the clues which suggested the diagnosis. Furthermore, the presence of Chiari’s triad of midthoracic pain, sentinel arterial haemorrhage, which is usually haematemesis, and final exsanguination after a symptom free interval was manifest in this case. Also, although not evidenced, placement of a cuffed endotracheal tube at the fistula sight was helpful in compressing and tamponading the fistula.

Competing interests None.

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