CASE REPORT

Aortoduodenal Fistula as an Unusual and Fatal Manifestation of Giant-cell Arteritis

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Introduction

Giant-cell arteritis is a focal, granulomatous vasculitis that is rarely found in patients younger than 50 years of age.¹ Patients with giant-cell arteritis of the temporal artery characteristically present with headache, jaw claudication or a painful temporal artery. However, giant-cell arteritis may involve all the medium sized or large blood vessels and therefore the symptomatology and the presentation of the disease is variable. The extracranial vessels are involved in about 13% of the patients suffering from giant-cell arteritis. We report a case of giant-cell arteritis presenting as an aortoduodenal fistula of the abdominal aorta.

Case Report

A 55-year-old man was admitted for evaluation of haematemesis associated with a 3 week history of dyspepsia. He had no history of abdominal, back, or joint pain, nor did he complain about fatigue, vision problems, or headache. He took no medication and his bowel movements were normal. Physical examination revealed a well-appearing white male, blood pressure of 140/50 mmHg and pulse rate of 62 per min. No abnormalities were found in the abdomen. Abdominal examination revealed no epigastric tenderness nor clinical manifestation of a dilated aorta. No fresh blood or melena was found on rectal examination. Laboratory results were as follows: ESR 14 mm/h, haemoglobin value 8.1 mmol/l (13.1 g/dl), haematocrit 42% with normal cell indices, white cell count 10.8 × 10⁹/l and platelet count 348 × 10⁹/l. The liverfunction tests and serum amylase were also normal. Coagulation studies yielded normal values for the bleeding time, prothrombin time and activated partial thromboplastin time. All other laboratory results were normal. Faecal occult blood was positive.

Gastroendoscopy revealed no obvious source of bleeding. Ultrasonography of the abdomen also showed no obvious abnormality. The abdominal aorta had a diameter of 3 cm. He was discharged on oral omeprazol 40 gm daily. Eight days later he was found in shock having passed large amounts of melena. Laboratory results revealed a drop in haemoglobin concentration to 5.5 mmol/l (8.9 g/dl). Gastroendoscopy showed blood in the distal part of the aorta.

Gut 1 The aortic wall next to the fistula with giant-cells and remnants of elastic fibres. HE, original magnification × 600. Reproduced here at 50%.

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duodenum. Once more no actual bleeding site was found. Before an emergency laparotomy could be performed, the patient became hypotensive and died as a result of exsanguination. At autopsy a fistula was found between the infrarenal aorta and the third part of the duodenum. No other fistulae were found. The aorta, with a diameter of 3.5 cm, was not aneurysmal. Macroscopically no atherosclerotic, inflammatory or obstructive manifestations were seen in the large abdominal and thoracic arteries (including the coronary vessels). Microscopic examination of the fistula revealed round cell and giant-cell infiltration of the media as well as disruption of the lamina elastica interna without granulomata (Fig. 1). No other localization of giant-cell arteritis was present, although the temporal arteries have not been examined separately.

Discussion

Primary aortoenteric fistulae are rare and their underlying etiologies are variable. The most common cause of a primary aortoduodenal fistula is atherosclerosis. Aortoduodenal fistulae have been described as a complication of gastrointestinal diseases such as a neoplasm, ulcers, cholelithiasis, diverticulitis, appendicitis, trauma and infectious aortitis. To our knowledge giant-cell arteritis, as a cause of aortoduodenal fistula, has not previously been reported. Giant-cell arteritis is an important manifestation of giant-cell arteritis and can be complicated by aortic arch syndrome, aortic valve regurgitation and even rupture of the aneurysmotic aorta. An aortopulmonary fistula has been documented1 but to our knowledge an aortoenteric fistula has never been described.

An elevated ESR is characteristic for giant-cell arteritis, however (as in our case) a normal ESR does not rule out the diagnosis. The course of the disease can be regarded as characteristic for an aortoduodenal fistula. Approximately 70% of the patients with an aortoduodenal fistula present with haematemesis or melena. The bleeding is characteristically intermittent, usually starting with a relatively short episode, the 'herald-bleeding', which is seldom fatal, only to be followed by a massive bleeding. The diagnosis is often made post mortem due to the fact that an aortoduodenal fistula is unlikely to be considered in a patient with haematemesis without a prior history of aortic surgery. The results of radiologic examination and ultrasonography are often limited. The primary role of a gastroduodenoscopy is to exclude other causes of gastrointestinal bleeding but cannot be regarded as complete if the distal part of the duodenum has not been visualised. If a fistula is visible or suspected, an immediate laparotomy should be performed. In patients with haematemesis and negative gastroduodenoscopy the possibility of an aortoduodenal fistula should always be considered.

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References


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