CASE REPORT

Ruptured inflammatory aortic aneurysm with aortoenteric fistula and infected with *Streptococcus pneumoniae*: a review of the literature

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Abstract We present a case report of a 65-year-old gentleman who presented with an aortoenteric fistula along with a review of the literature. He was found, in addition, to have an inflammatory infrarenal aortic aneurysm with a posterior rupture. Cultures of the aneurysm sac confirmed a *Streptococcus pneumoniae* infection. The patient had previously presented with pneumonia. Antibody testing revealed an isolated pneumococcal IgG deficiency. The case demonstrates the diverse pathologies associated with aortic aneurysms and a need to be vigilant and occasionally expect the unusual.

Keywords Abdominal aorta · Aneurysm · Inflammatory aneurysm · Mycotic · *Streptococcus pneumoniae* · Fistula · Aortoenteric

Introduction

Primary aortoenteric fistulas have become increasingly rare with the modern aggressive management of aortic aneurysms. The most common aetiology is atherosclerosis, but they can also be secondary to infection, trauma, radiation, neoplasia and foreign bodies [1–4]. The commonest presentation is haematemesis, melaena and abdominal pain. The addition of lumbar or back pain, a clinically palpable abdominal mass, or the presence of a known aneurysm should increase the clinical suspicion of an aortoenteric fistula [1–4].

The most common site for fistula formation is the third part of the duodenum. This is due to its relatively fixed position as compared with the rest of the bowel, and the close relationship of this part of the duodenum to the aorta. Fistula formation has however been documented from stomach to rectum [1–5]. We present a case that demonstrates the difficulties in diagnosis and management, and present a current review of the literature.

Case report

A 65-year-old male presented to our emergency department, collapsed and shocked with extensive haematemesis. On examination, he demonstrated marked abdomen tenderness and had a palpable pulsatile abdominal mass. He had recently been discharged from another hospital having been complaining of a two-month history of increasing abdominal and back pain. This was at first thought to be renal colic. Investigations had included an unremarkable ultrasound scan of his abdomen and pelvis. He had also undergone a bone scan to investigate chronic back pain. This was also negative. His other medical history of significance was an admission for pneumonia eight months previously.

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He underwent immediate resuscitation with blood and crystalloids and was transferred to the operating theatre. Laparotomy revealed a 4.5 cm diameter inflammatory, infra-renal aortic aneurysm with a fistula extending into the proximal jejunum. The jejunum was dissected off the aneurysm wall, thus isolating the fistula and exposing the aneurysm sac. The aneurysm neck and both iliac arteries were then clamped. The jejunal fistula was repaired in two layers with absorbable sutures. On opening the aneurysm sac, a posterior rupture was also noted with a retroperitoneal haematoma. The aneurysm was repaired in standard fashion with a rifampicin soaked, 16×8 bifurcated graft (Sulzer Vascutek) via an inlay technique, to the common iliac arteries.

Immediately following the repair, no pulse was felt in the right common iliac artery. A thrombectomy was performed followed by a Dacron patch angioplasty; flow was restored. The aneurysm wall, fistula and clot were sent for histology and microbiology. The patient was transferred to the intensive care unit, stable, intubated, sedated, but requiring inotropic support with adrenaline.

Two days later, histology confirmed that the aneurysm was inflammatory in nature and was also staining positive for gram-positive rods (Fig. 1). Microbiology subsequently reported that the fistula, aneurysm, and clot present inside all cultured a *Streptococcus pneumoniae* sensitive to amoxycillin. This was added to his ongoing regime of cefu-

roxime and metronidazole. The *Streptococcus pneumoniae* was subsequently demonstrated to be sero-group 23, sub type 23F (Central Public Health Laboratory, London). Further blood samples were sent for immunological screening of the patient.

He remained stable and well for the next 6 days. He was gradually weaned off inotropic support, woken up, extubated and started with feed. However, on day 7 he developed an ileus. His urine output decreased and he developed signs of septicaemia. A laparotomy revealed faeco-purulent fluid in his abdomen and an ischaemic bowel from mid-transverse to proximal sigmoid colon, with a perforation at the sigmoid apex. He underwent a left hemicolectomy and end colostomy.

He continued to make progress until again becoming septic on day 12 post aneurysm repair. An abdominal CT scan revealed no significant pathology. Blood cultures grew gram-positive rods for which vancomycin was prescribed. At this stage immunology results indicated that he had isolated low antibody titres to *Streptococcus pneumoniae*. He was given the pneumovax vaccination.

Following seven days treatment, the vancomycin was stopped and co-amoxiclav started to protect the graft in view of the abdominal sepsis. He remained in a stable condition requiring only respiratory support in the intensive care unit until day 36 when his oxygen requirements increased and sputum cultures revealed a methicillin resis-

Fig. 1 Gram stain of aortic wall showing gram-positive cocci colonies (x 400)

tant *Staphylococcus aureus* (MRSA). Vancomycin was re-started.

Two days later it was noted that his right arm had become swollen. Duplex ultrasound showed his axillary veins to be patent. This progressed into an erythematous and bullous rash that began to spread to his trunk and legs. This was biopsied and found to be toxic epidermal necrolysis, presumably secondary to vancomycin. His antibiotics were stopped, he was given alphaglobulin and priobrane dressings. Despite this his condition deteriorated and he died on day 40.

Discussion

Mycotic aneurysms of the aorta are rare [6]. Bacterial infections account for 18% with *Streptococcus* spp. accounting for approximately 8% of these cases [7]. Gram-positive organisms are responsible for 60% of infected aneurysms with *Staphylococcus aureus* accounting for 46% of cases. Gram-negative organisms have greater virulence with a death rate of 72% as compared to 25% for gram-positive organisms [7]. Since 1966 there have been 9 further cases of *Streptococcus* infected aneurysms reported in the English literature [7].

Aortoenteric fistulae are also rare. Most are of the secondary variety and follow vascular surgery. They most commonly occur between the aorta and duodenum although many other parts of the bowel have been reported. The presence of an infected aorta leading to an aortoenteric fistula and spontaneous rupture is reported in the literature on only one previous occasion. In that case the organism was assumed to be *E. Coli* [8].

Previously *Streptococcal* endocarditis was regularly reported [9]. With modern antibiotics the incidence has fallen. Pneumococcal lung infection can lead to a bacteraemia that may seed infection to distant sites, in this case the abdominal aorta [9]. Contiguous spread to the aorta from a diaphragmatic abscess has also been reported [7]. Bacteria are known to produce and secrete bacterial proteinases [10]. This response is independent of matrix metalloproteinases, the accepted culprit behind the dilatation of non-infected abdominal aortas [11]. Bacterial proteinases can degrade the matrix and produce a rapidly expanding aneurysm. This aneurysm had ruptured at 4.5 cm, for a non-infected aneurysm one would expect a rupture rate of less than 2% per year from the UK small aneurysm trial [12]. In the presence of infection, matrix degradation and rapid expansion they may rupture at a smaller diameter. Clinical examination and abdominal ultrasound failed to demonstrate an aneurysm 2 months prior to its rupture.

This patient had received appropriate antibiotic cover for his pneumonia and at the time of surgery. His cultured aneurysm wall grew type 23-*Streptococcus pneumoniae*. Subsequent antibody testing demonstrated only an isolated

deficiency of *Streptococcal* antibodies in this patient. This may have produced and participated in *Streptococcal* aortitis in our patient and help in explaining the virulence.

Sir Astley Cooper first reported and described an aortoenteric fistula in a post mortem case in 1829 [13]. Since then 253 cases have been documented in the literature [14]. The incidence of primary aortoenteric fistula was reported as 7 in 10,000 consecutive cases in a post mortem study [15], and 2 of 3,000 aortic aneurysm resections [16]. Most occur secondary to atherosclerotic disease (73.5%) with infection being the underlying cause in 10.7% [14].

Aortoenteric fistulas have an overall mortality of 71% from the 253 documented cases [14]. One hundred and sixteen of the cases (46%) were operated on, with a 30-day mortality of 37% [14]. They generally present with pain (32%), haematemesis (64%), a mass (25%) or hypovolemia and shock (5%) [4]. The duodenum is the site of the fistula in 86.4% of cases [14]. Treatment options include debridement and toilet of the infected aorta and surrounding tissues with either an extra-anatomical or an inlay graft, or suture and patch repair. Most cases (81%) have been repaired with an inlay graft [14]. Antibiotic or silver impregnated grafts may offer an additional advantage.

Inflammatory aneurysms have remained somewhat of an enigma. Walker et al first coined the term 'inflammatory' in 1972 to describe the 10% of aneurysms in their series with a 'thick, firm, smooth wall, which is shiny white in appearance, with dense fibrosis which extends to involve adjacent structures' [17]. In the elective setting the diagnosis of an inflammatory aneurysm is often made preoperatively by CT scan as thickening of the adventitia and media of the wall can be seen. Clinically the triad of a thickened aneurysm wall, extensive fibrosis and dense adhesions confirms the diagnosis. They tend to present in a slightly younger age group (64–66 years) with a male to female ratio of 80% to 20% [18]. The aetiology remains unknown. Several theories have been postulated such as infection, lymphatic stasis due to the compression from the aneurysm, and an autoimmune response [17, 19]. To date none have been proven. Our report may add some weight to the infection theory.

Inflammatory aneurysms tend to produce more symptoms than their non-inflammatory counterparts. Fever, malaise, weight loss (20%) and tenderness (22%) are common. A raised ESR may be seen in the blood profile. Renal function tests may show deterioration as the ureters may be involved in the inflammatory process (18.5%) producing an obstructive nephropathy [18]. Reports suggest that once the aneurysm is repaired the fibrosis and inflammatory process will resolve.

Toxic epidermal necrolysis (TEN) is almost always a severe drug-induced reaction and does not appear to be caused by a circulating bacterial toxin [20]. TEN causes a full-thickness epidermal necrosis that is fatal in 25% of cases [21]. Initial symptoms that precede epidermal necrosis include fever, conjunctivitis, pharyngitis and pruritus

[20]. These signs can resemble an upper respiratory tract infection. Management of these patients is similar to a patient with severe burns and includes careful protection of exposed dermis, fluids and nutritional support.

We postulate that the pneumococcal infection started this sequence of events possibly by seeding on an atherosclerotic plaque within the aorta. This then led to the development of an inflammatory aneurysm with the jejunum becoming involved and producing a fistula. We had considered whether a generalised immunodeficiency was the underlying causal factor in this case. This was not demonstrated, however, the patient had an isolated pneumococcal immunoglobulin deficiency. Was this the cause for the infection or did it arise as a result of the infection? With a retrospective perspective, it is always difficult to be sure.

This case demonstrates a rare condition. The patient presented 6 months following pneumonia with a rapidly expanding and ruptured aortic aneurysm and the presence of an aortoenteric fistula. It demonstrates the diverse pathologies associated with aortic aneurysms and a need to be vigilant and occasionally expect the unusual.

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