JNDS Journal of the Nuffield Department of Surgical Sciences

Case Study

The challenges of diagnosing and managing a ruptured abdominal aortic aneurysm with primary aortoduodenal fistula.

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Keywords:
Aortoduodenal fistula, aortoenteric fistula, abdominal aortic aneurism, vascular surgery.

Summary
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This is a detailed analysis of a very rare, life-threatening condition. The vast majority of aorto-enteric fistulae occur in patients who have had previous aortic surgery. The presentation of a primary aorto-enteric fistula associated with a ruptured abdominal aortic aneurysm is a true singularity that provides a unique, gargantuan challenge for any clinician. This case report provides a detailed account of this fascinating case, together with a thorough overview of the current evidence base for potential diagnostic and management strategies.

Introduction

When William Osler asserted, "There is no disease more conducive to clinical humility than aneurysm of the aorta."1 he was referring to the heterogeneity of the clinical presentation, and the catastrophic consequences of misdiagnosis. The development of a primary aortoduodenal fistula from the abdominal aortic aneurysm (AAA) serves only to complicate every aspect of the illness, broadening the variability in clinical presentation, complicating the surgical treatment, and adding a complex microbiological management aspect. The resulting disease is fatal when untreated or treated medically, and even if surgical treatment is begun in time, there is a high risk of intraoperative and perioperative mortality and serious long-term complications.

The case of X, a patient who underwent an emergency open repair for a ruptured AAA complicated by a primary aortoduodenal fistula provides an interesting platform to discuss the presentation and management of this rare disorder, highlighting complicating factors such as the heterogeneity of presentation, lack of definitive imaging techniques, and lack of evidence regarding surgical and microbial management.

Patient X Case History

Patient X is a 68 year old man who presented in December 2016 with a ruptured abdominal aortic aneurysm with a primary aortoduodenal fistula. When he presented, he had a 4 day history of malaise, loss of appetite and back pain before collapsing. He was brought in as an emergency case and after diagnosis by CT, underwent an open "trouser graft" repair with an ovine omniflow biograft. During the operation, they removed a large clot from the aorta which tested positive for streptococcus gordonii and streptococcus anginosus. He was treated post-surgically with IV co-amoxiclav for 28 days before being stepped down to IV ceftriaxone and oral co-amoxiclav.

In March 2017, shortly after the step down from co-amoxiclav, X presented with sepsis, and a, and during this visit based on CT findings suggestive of proximal pseudoaneurysm and dehiscence of the proximal anastomosis between the biograft and right common iliac artery, complicated by graft infection he underwent extensive graft revision surgery. The surgeons performed a right renal chimney repair, aorto-uni-iliac stenting, right femorofemoral crossover and a right common femoral endarterectomy. After this surgery, he was sent home, on IV ceftriaxone which were stepped down to lifelong prophylactic co-amoxiclav.

X presented again in December 2018 with sepsis, with blood cultures positive for streptococcus adjacens and enterobacter cloacae, and CT-A showing gas in perigraft area. He was treated initially with IV ceftriaxone and oral metronidazole which was stepped up to IV ertapenem and gentamicin. He was also given left iliac angioplasty and stenting to improve perfusion of his lower limbs which was shown by angiography to be lacking. During this admission, the possibility of vascular explant surgery to remove the graft was discussed with the patient, who turned it down based upon the high mortality rates of the operation.

In January 2020, X presented again with sepsis, and acute kidney injury as a result of septic emboli from the infected graft infarcting the right renal artery. Cultures grew actinomyces and enterococcus faecium X was managed medically, initially with IV meropenem and vancomycin and then sent home on IV ertapenem and teicoplanin. X understands that he is unlikely to regain significant function in his right kidney, and remains on lifelong prophylactic co-amoxiclav.

Patient X provided written informed consent for his case to be written up as a case study.

Aortoduodenal fistulas - an overview

Primary aortoduodenal fistulas are an abnormal connection between the endothelium of the aorta and the endothelium of the duodenum, formed without prior aortic reconstructive surgery. First described in 1818 by Sir Astley Cooper[2], they are an exceptionally rare condition, with an annual incidence of 0.007 per million[3]. As of 2014 there were around 200 cases of primary aortoduodenal fistulas in the literature. Other aortoenteric fistulas also occur - there are around 150 described in the literature, but due to the anatomical proximity of the third and fourth segments of the duodenum to the aorta, these are the most commonly involved sites[4].

Aortoenteric fistulas are thought to result from a large aneurysm eroding into the bowel wall[5] - atherosclerotic in around 73% of cases and traumatic or mycotic in 26%. The remaining 1% were due to a variety of causes including malignancy, radiation, ulcers gallstones, diverticulitis and cystic medial necrosis[6]. There is some evidence from animal experiments that factors contributing to their development may include a combination of mechanical factors and infection and inflammation though results conflict as to which factor is more important[7,8]. There is a body of evidence documenting cases where a number of different organisms - salmonella and klebsiella most commonly but also, haemophilus, E.Coli, clostridia, TB, syphilis, mycosis, streptococci, and staphylococci[6,9] - causing septic aortitis have resulted in formation of aneurysms and pseudoaneurysms, and some in which these aneurysms then eroded into the gut. The obvious flaw in this body of evidence however being that the aortoenteric fistulas having been identified having already formed, meaning that no causation can be assessed however since since septic aortitis merits aggressive antimicrobial therapy when detected[10], this flaw is likely to be insurmountable in human studies.

The risk factors for primary aortoenteric fistulas are relatively poorly characterised with the exception of the existence of an AAA. Certain infections - syphilis and tuberculosis - as well as collagen vascular disease and previous mycotic infarction are risk factors[11]. Tareen and Schroeder[12] published a review of 44 cases finding a male to female ratio of 4:1, and an average age of presentation at age 63 (range 23-82). Given the paucity of cases for analysis it is hard to distinguish whether there are risk factors for aortoenteric fistulas themselves, or simply increase risk of fistulas by increasing risk of AAAs, which are by far the largest risk factor. Risk factors for AAAs can be broadly grouped into atherosclerosis, history or family history of vascular disease, hypercholesterolaemia, hypertension, male sex, obesity, age, and tobacco use[13].

Clinical Presentation and Diagnosis

Primary fistulae present most commonly with GI bleeding (~80% of cases), abdominal pain (32%) and a pulsatile mass (25%). The GI bleeding may be intermittent[14] in which case it may be a "herald bleed" for massive GI haemorrhage. Other, less common symptoms include back pain, melena, fever, sepsis, and shock.

Patient X presented to hospital with sudden onset back pain, and collapse due to hypovolemic shock, meaning that he had urgently CT-A on the basis that a ruptured AAA was suspected, and this led to the diagnosis of his aortoenteric fistula. While patient X's aortoenteric fistula was detected due to his presenting with AAA rupture, a patient presenting prior to rupture may have subtle and non specific symptomatology. The presenting symptoms are relatively non-specific and may be consistent with a number of other conditions which are also medical emergencies. GI hemorrhage could equally represent ruptured GI ulcers or varices, malignancy, haemobilia, or Dieulafoy lesion rupture. Pain (which depending on site of rupture this may be felt in the back, abdomen, chest or groin) has any number of differential diagnoses, and those relating to infection are entirely non specific for aortoenteric fistula. The diagnosis, therefore, depends almost entirely on imaging, exploratory surgery, or post-mortem, and without surgical treatment, the mortality rate nears 100%[6] and for these reasons it is important that the clinical index of suspicion is high so that the appropriate investigations can be carried out.

The imaging modalities most useful for the detection of aortoenteric fistulas are CT-angiography (CTA), oesophagogastroduodenoscopy (EGD), and arteriography[5].

In an acute setting CTA is likely to be by far the best method of making the diagnosis in a time-sensitive manner. One of the most suggestive signs of an aortoenteric fistula is pneumoretroperitoneum[3], however this is not a specific finding and is therefore only likely to lead to diagnosis if the index of suspicion is high. Extravasation of arterial contrast in the gut lumen, a pathognomonic finding, was only present in 1/3 cases in a study considering both primary and secondary fistulae[15]. Other highly suggestive findings include loss of delineation of the wall of the aneurysm or the fat lying between the aorta and duodenum, however much of the literature on the matter is derived from and therefore more relevant to secondary fistulas than primary[3]. Overall the sensitivity of CTA is estimated to be between 40-90%, and the specificity 33-100%[16], and therefore absence of suspicious signs on CTA certainly cannot exclude aortoenteric fistula[5].

EGD and arteriography are more invasive, slower to perform, and less readily available, and require the patient to be haemodynamically stable. EGD carries the additional risk of physical trauma to the AAA or fistula, as well as the risk of dislodging an aortic thrombus. It is often used first line if the patient presents with upper GI bleeding[14] as it is useful in ruling out other causes of upper GI bleeding. It may reveal associated pathology[17] or even highly suggestive findings such as a pulsatile mass in the duodenum18 but while there are many reports in the literature of EGD being diagnostic for secondary aortoenteric fistulas, diagnosis of a primary fistula usually requires confirmation from a second imaging modality if EGD is the initial approach[19,20]. A negative EGD is certainly not sufficient to rule out an aortoenteric fistula[5,6]. Arteriography, like CT-A, may show pathognomic extravasation of contrast into the gut lumen but is often non-diagnostic with one review showing that in 15 cases of aortoenteric fistula aortic angiography was only positive in 2[12]. Based on this, arteriography should not be considered as an initial approach in suspected primary aortoenteric fistulas.

Multi-detector CT and MRI may be useful alternatives - especially in renal patients where MRI can avoid exposure to IV contrast[14]. There is some suggestion in the literature that ultrasonography may also be useful in these patients[5].

Treatment Strategies and Outcomes

Untreated, the overall mortality of aortoenteric fistulas is almost absolute[6]. Treatment is either urgent, or emergent, surgery as evidence suggests that the survival rate is inversely related to the delay between the onset of GI bleeding and surgical intervention[5]. The repair of an aortoenteric fistula presents a significant

technical challenge and mortality rates even in relatively uncomplicated cases are around 30%[5]. The rarity of the disease has led to a paucity of high quality large or long term studies meaning that the choice of surgical approach remains controversial[21] and will of course depend on the presentation and patient. Extraanatomic bypass with aortic ligation has been the historical gold standard but this is being increasingly challenged by various in situ methods of maintaining perfusion[22]. Some consider a multiple step approach – for example patching the aorta and repairing the duodenum and scheduling elective AAA repair - to put the patient at lower risk of infection than a single definitive surgery[3].

There is no significant difference in overall mortality between open and endovascular repair in cases uncomplicated by rupture and where the aneurysms are non-inflammatory[23]. Endovascular repair is associated with better short-term outcomes with lower risk of the key post-operative morbidities: mediastinal abscess, acute renal failure, and bowel obstruction, as well as infection in the short term, but this balanced by increased incidence of long-term infective complications[5]. Where the patient is fit, however, there are a number of advantages to open surgery. Open surgery can be necessary to confirm the diagnosis, allows repair of the bowel defect, allows more effective source control if the site is infected[5], and is associated with a lower risk of recurrence, and recurrent postoperative hemorrhage and infection in the long term[26].

Bowel repair, including interposition of the omentum between the aorta and duodenum, is a vital component of a successful surgery as the most common cause of death post operatively is recurrence of the fistula, which is significantly more likely if the patient is treated with a duodenorrhaphy without interposition[4]. Delayed enteric repair following endovascular repair is emerging as a treatment option with some early studies demonstrating it to be promising[4,24,25]. In a report of 2 patients treated endovascularly, both had good outcomes but required operative revision 9 and 16 months later respectively[26]. Based on these reports, it seems likely that an approach may be developed based on this idea that will go on to become the new gold standard for aortoduodenal fistula repair.

Open surgery again is a better treatment in cases where there is significant contamination. In cases of severe retroperitonitis, mycotic fistulas[6], and gross contamination[21], the surgery of choice is extensive debridement with an extra anatomic bypass graft. Patients who present with evidence of infection have worse outcomes with endovascular surgery[27] although postoperative mortality rates for open repair of primary aortoenteric fistulas associated with infected AAAs are still in the region of 50%.

In cases of rupture or significant GI bleeding, open surgery may be preferred to avoid the risk of failed endovascular intervention meaning an open surgery need then be attempted with the patient's physiological reserve further reduced[3]. If the patient is already unfit for open surgery, however, endovascular repair may be used as a stopgap to stabilise the patient for later open surgical repair of the fistula[5,14].

Endovascular repair and antibiotic therapy alone are rarely an appropriate treatment. The operation may allow aortoenteric fistulas to heal in select patients, especially those presenting without symptoms of infection[5] but as a general rule the high rate of recurrence, recurrent postoperative hemorrhage and recurrent postoperative infectious complications it should be considered as a temporary measure to optimize the patient for open repair[26]. One caveat to this is in palliative patients, where endovascular repair alone may be useful[5] allowing for relatively short hospital stays and greater probability of discharge home[28].

Whatever the procedure attempted, it is crucial that intraoperative cultures should be taken in order to tailor therapy to the sensitivities of the bacteria present, and even in case the culture is negative prophylactic antibiotics should be given[6] along with antifungal cover[24]. Indeed, broad spectrum antibiotic therapy covering at least gram positive, gram negative, and enteric pathogens should be initiated as soon as the diagnosis is suspected. Post surgically this can be tailored to the bacteria cultured during the operation and should continue for a minimum of 6 weeks if cultures are positive. There is dispute in the literature about perioperative antibiotics with negative intraoperative cultures: with suggestions of a minimum of 1 week[6,21] and a minimum of 65, however this is insufficient evidence to conclude either way. One option is to track erythrocyte sedimentation rate and C-reactive protein levels and adjust the length of treatment based upon these results[24]. Once this period ends, patients should remain on prophylactic antibiotic therapy for life[5].

Discussion

Over the course of this discussion of primary aortoduodenal fistulas as a complication of AAAs, the most significant points are the heterogeneity in clinical presentation and findings on imaging and the lack of evidence regarding management.

Patient X presented as an emergency with shock and symptoms of a AAA meaning that he was treated as an emergency and therefore received the rapid surgical intervention required but those presenting with less characteristic symptoms are at risk of fatal exsanguination either through GI haemorrhage or aortic rupture. This risk is increased by the fact that none of the 3 standard investigations - CT-A, EGD, and angiography - are able to definitively exclude the diagnosis. For this reason, it is vital to maintain a high index of clinical suspicion even though aortoduodenal fistulas are exceptionally rare without a previous history of aortic surgery.

Patient X had a successful operation, but has gone on to require multiple revisions, and significant morbidity in the form of recurrent sepsis and AKI. He may have benefitted from a partial duodenal resection either during the initial surgery or at a later date to reduce the risk of recurrent graft infections. He also would likely have benefitted from a better body of research on pre- and peri-operative microbial management, which are still handled ad-lib due to a lack of scientific consensus. One key area which remains to be delineated is whether the management of his ongoing graft infection should be any different having resulted from an aortoduodenal fistula.

The surgical approach to aortoduodenal fistulas is developing over time, but it seems likely it may evolve to favor multiple step operations where possible, combining both the good short term outcomes of endovascular surgery, and the reduction in the risk of long term complications in open surgery, as well as ensuring the haemodynamic stability of the patient before open surgery is attempted. Of course, in emergency cases or where the diagnosis is not clear from imaging, open surgery will still likely have a role as the primary approach.

Conflicts of interest
None.

Funding
None.

Consent
The patient has consented to the publication of this case study.

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