Primary aortoenteric fistula caused by an infected abdominal aortic aneurysm with *Mycobacterium Avium* Complex in an HIV patient

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Abstract

Primary aortoenteric fistula (PAEF) is a rare but complex clinical entity requiring multimodality approach for diagnosis and treatment. We report the first ever case of upper gastrointestinal (UGI) hemorrhage caused by an aortoenteric fistula (AEF) secondary to *mycobacterium avium* complex (MAC) in a patient with human immunodeficiency virus (HIV) infection. Esophagogastroduodenoscopy (EGD) showed an ulcer in the distal duodenum and a computed tomography (CT) scan confirmed a contained abdominal aortic aneurysm (AAA) rupture with an aorto-enteric fistula communicating with the third portion of the duodenum. Emergent surgery was undertaken which was lifesaving. A high index of suspicion, early diagnosis and prompt surgical intervention are crucial for survival of patient with PAEF. (Acta gastroenterol. belg., **2010**, 73, **280-283**).

Key words : Primary aortoenteric fistula, *mycobacterium avium* complex, upper GI bleeding.

Case report

A 48 year-old-male presented to the emergency room with one episode of hematemesis and hematochezia. Prior history included HIV which was diagnosed one year back for which he was taking highly active antiretroviral therapy (HAART) and sulfamethoxazole/trimethoprim prophylaxis. His CD4 cell count was 146 cells/µL and viral load was less than 50 copies/mL. Other medical problems included an abdominal aortic aneurysm. A CT scan 1 month back showed a size of 4 cm. Physical examination revealed a blood pressure of 80/60 mm Hg and a heart rate of 116 beats per minute. The abdomen was soft and non-tender. The liver and spleen were not felt, and no abdominal mass was appreciated. The peripheral pulses were intact. Laboratory tests revealed a hemoglobin of 12.9 g/dL (13.8-17.2 g/dL) and a hematocrit of 38.6% (44-51%). After initial resuscitation, an emergent EGD was performed. The esophagus and stomach were normal. However, a 3 cm ulcer was noted in the distal duodenum (Fig. 1). A CT scan of the abdomen revealed a contained AAA rupture of the infraarenal portion of the abdominal aorta with an AEF communicating with the third portion of the duodenum (Fig. 2). Broad spectrum antibiotics including levofloxacin and vancomycin were started and an emergent surgical exploration was undertaken. At laparotomy, a mid-line incision was made extending from the xiphoid process all the way down to pubic symphysis. On entering the retroperitoneum, an aortoenteric fistula was found between the third part of the duodenum and the proximal infrarenal aortic aneurysm. Infrarenal proximal control above the aorto-enteric fistula and the distal control at the common iliacs was obtained. After that, the diseased segment of the aorta was resected and primary repair of the AAA with axillo-bifemoral Dacron bypass grafting was done. The duodenum was closed in two layers and an omentopexy was performed between the aorta stump and the duodenum. Gram staining was negative but microscopy using Ziehl-Neelsen staining showed acid-fast bacilli (AFB) with a beaded appearance (Fig. 3). Histopathology revealed non-caseating granulomas with scattered foci of AFB in the resected aortic aneurysm wall (Fig. 4). Species identification using DNA probes distinguished *mycobacterium avium*

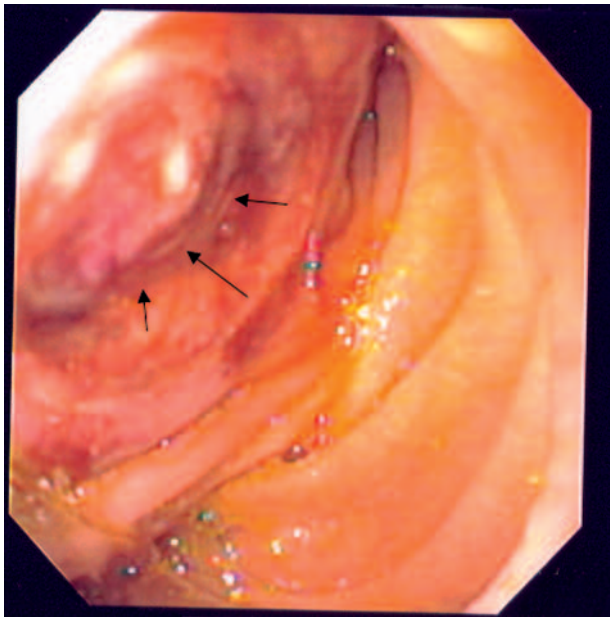

Fig. 1. — Endoscopic view showing a large ulcer in the distal duodenum (arrows).

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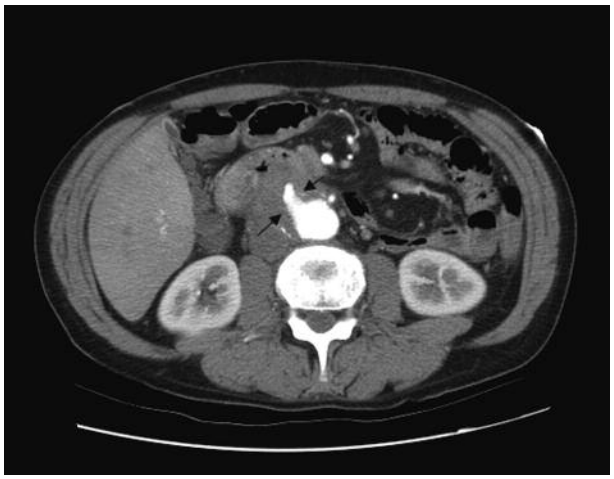

Fig. 2. — CT scan of abdomen showing contained perforation of the infrararenal portion of the abdominal aorta with the aortoenteric fistula (arrows).

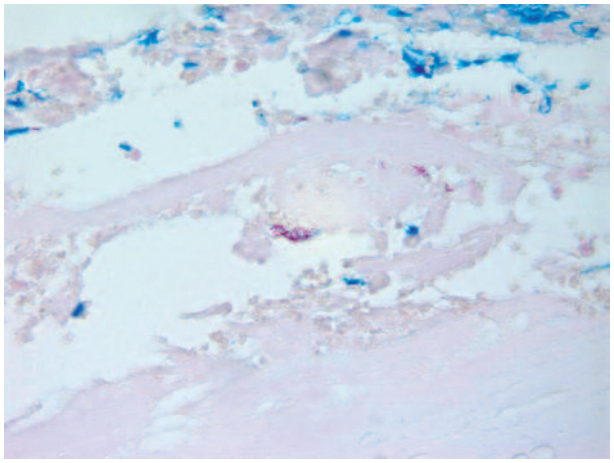

Fig. 3. — Microscopy using showing acid-fast bacilli with a beaded appearance in the fibrinous debris (Ziehl-Neelsen stain, 400×).

complex (MAC) from *M. tuberculosis*. Prior to resuming oral intake, he was treated with intravenous azithromycin, levofloxacin and rifampin. He was able to tolerate oral feeding on the fifth post-operative day. His HAART therapy was restarted and the antibiotics were switched to oral azithromycin, rifabutin and ethambutol. The patient had an uneventful post-operative course and was discharged 15 days after the admission. At 12-month follow-up, his viral load had remained undetectable for the preceding six months and his median CD4 cell counts has been 240 cells/μL, so the maintenance therapy for the MAC infection was discontinued. From there on he was continued with HAART and sulfamethoxazole/trimethoprim prophylaxis. He has so far completed 2-year follow-up ; is alive and doing well.

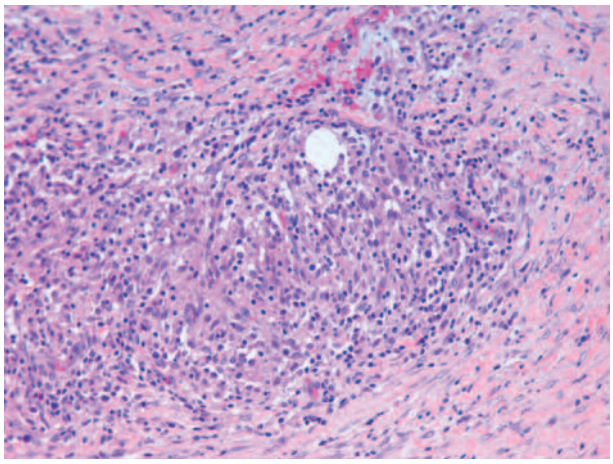

Fig. 4. — Histopathology showing non-caseating granulomas with scattered foci of acid-fast bacilli in the resected aortic aneurysm wall (haematoxylin and eosin stain, 100×).

Discussion

Aortoenteric fistula is classified as primary and secondary with the latter being more common with a reported incidence rate of 1.5%-4% and usually is an ominous complication of aorto-iliac reconstructive surgery (1). On the other hand, PAEF is a rare but life-threatening cause of acute UGI bleeding. First described by Cooper (2) in 1832 ; it is defined as a spontaneous development of a connection between the native aorta and the GI tract. The reported incidence is 0.04%-0.07% in large autopsy series (3). One large review has reported a 2.8 : 1 male preponderance with a mean age for diagnosis of 64 years (4).

Atherosclerosis accounts for more than two thirds of reported causes. Other uncommon causes include trauma, infections, radiation, metastasis, pancreatic carcinoma, ulcers, gallstones, diverticulitis and appendicitis (4-6). Because of the close approximation and fixed nature of the duodenum, it is thought that the expanding nature of the AAA causes inflammation and pressure necrosis, resulting in eventual fistulization over time (7). Infectious agents involved in PAEF are most commonly *salmonella* or *klebsiella* ; however, *treponema pallidum*, *E. faecalis*, *mycobacterium tuberculosis*, *clostridium septicum*, l*actobacillus*, *coxiella burnetii*, mycosis have all been reported (1,8). We report the first case in literature of MAC causing PAEF presenting with UGI bleeding. In patients infected with HIV, MAC infection usually present as disseminated disease although localized forms of MAC are being reported with widespread use of HAART.

The characteristic bleeding for an aortoenteric fistula is similar to other forms of gastrointestinal bleeding. The presentation is often subtle, with intermittent “herald bleeding” which usually precedes hypovolemic shock by a variable period of time, making it important to recognize this event when it occurs. Fifty percent have longer

than 24 hours between the herald bleed and their exsanguinations, while 29 percent go more than 1 week (6) Conversely, 30 percent of patients with a PAEF will experience a massive bleed within six hours following a herald bleed. Other signs and symptoms may include abdominal pain, back pain, pulsatile abdominal mass, fever and sepsis (9,10). A high index of suspicion is critical for making a successful diagnosis, which may then be evaluated by a CT imaging with contrast (11). In our case, the endoscopy showed the large ulcer in distal duodenum and the CT scan confirmed the contained AAA rupture with the AEF.

Surgery is the mainstay of treatment for PAEF. The surgical approach for primary AEF should take into consideration the cause of the primary AEF, degree of retroperitoneal contamination, and prevention of future AEF (12,13). Surgical repair of the aortic aneurysm and fistula is the standard treatment.

Many fistulas are discovered only during an exploratory laparotomy and under these circumstances an option is to insert a graft following discontinuation and closure of fistula. If possible, a portion of omentum is wrapped around the vascular anastomosis and the overlying intestines. Results have been surprisingly constant over several decades with mortality rates of between 30 and 40 percent. An alternative treatment is to disconnect the fistula followed by oversewing of the aorta and axillobifemoral bypass. However, this approach is thought to be inferior and should be reserved for patients with extensive local sepsis and sever peritonitis (14,15). Closure of the gastrointestinal defect as a stand-alone treatment has dismal results because of ensuring sepsis and multiple organ failure and has been abandoned.

Antibiotics should be administered before operation if a PAEF is suspected. If the fistula is treated by an open procedure, results of preoperative cultures may allow for adjustment in the postoperative period. Antibiotic treatment should be provided for at least 1 week following negative cultures ; systemic antibiotic treatment is continued for 4-6 weeks if cultures are positive (13). For AEF secondary to MAC, Combination drug therapy is necessary for rapid clearance of MAC from the bloodstream and to decrease the risk of drug resistance (16). Agents with MAC activity include clarithromycin, azithromycin, ethambutol, rifabutin, amikacin, streptomycin, and fluoroquinolones. The appropriate duration of MAC therapy and the duration of immune reconstitution before stopping treatment have not been determined. However, United States Public Health Service guidelines state that at least 12 months of therapy and six months of immune reconstitution may be reasonable parameters (17).

Endovascular abdominal aortic aneurysm (EVAR) repair has an established role in elective management of abdominal aortic aneurysms (AAA). However, the application of EVAR to ruptured AAAs (rAAA) is evolving (18). Although EVAR has been utilized to manage rAAA for greater than ten years, to-date no randomized study has been completed to confirm superiority over traditional open surgical repair. One large retrospective review from a tertiary referral centre has reported an overall mortality of patients treated for ruptured aortic aneurysm of 25.2%, with an open repair mortality of 27.2%, and EVAR mortality of 20% (19).

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