

# A Unique Clinical Presentation of Primary Aortoenteric Fistula

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## Abstract

Primary aortoenteric fistula is a rare but fatal cause of gastrointestinal bleeding. The diagnosis of primary aortoenteric fistula is difficult to make and is usually accompanied by a very high level of clinical suspicion. In the context of a known abdominal aortic aneurysm it is reasonable to have a high index of clinical suspicion of aortoenteric fistula and it should also be included in the differential diagnosis with low back pain and a palpable midline abdominal mass in a haemodynamically stable patient. We present the case of a 59 year old man with no past history of abdominal aortic aneurysm presented with lower back and periumbilical pain. Initial misdiagnosis led to a delay in treatment and the patient succumbed to the illness.

## INTRODUCTION

A primary aortoenteric fistula (PAEF) is a rare and life-threatening cause of massive gastrointestinal bleeding [1]. It is the result of a progressive erosion of abdominal aortic aneurysm through a segment of the intestines, most commonly the duodenum, resulting in a direct communication between the aorta and the gastrointestinal system, mostly with the third portion of the duodenum [2]. This differs from a secondary aortoenteric fistula, which appears more frequently and originates from a formerly prosthetic abdominal aortic vascular graft [1, 3, 4]. The incidence of PAEF has been reported to be 0.04–0.07% in large autopsy series [4, 5]. Arteriosclerosis is the main cause of PAEF, Gad et al [6] reported 73% of PAEF were from atherosclerotic aneurysms and 26% were from traumatic or mycotic aneurysms [7, 8], other proposed theories for the formation of a PAEF are direct wear, inflammatory destruction triggered by infection, radiation, metastases, pancreatic carcinoma, ulcers, gallstones, diverticulitis, appendicitis, and cystic medial necrosis [7, 9]. The most common infectious agents involved in mycotic aneurysms are Salmonella or Klebsiella; however, tuberculosis, syphilis, mycosis, Staphylococcus, and Streptococcus can also be responsible [7, 9].

The diagnosis of PAEF is usually difficult because the classic triad of abdominal pain, gastrointestinal bleeding and pulsatile abdominal mass is present only in a minority of the cases [1, 3, 4, 5]. Initial herald bleeding, manifested by

haematemesis is followed by massive bleeding and exsanguination in the majority of patients; therefore, a high index of suspicion is needed to establish the diagnosis of an aortoduodenal fistula [3, 5].

We present the case of a 59 year old man who initially presented with lower back and periumbilical pain, no past history of abdominal aortic aneurysm, no history of haematemesis and haemodynamically stable at admission. Later on he developed severe haematemesis. Bearing this clinical presentation in mind in future could avoid a misleading initial diagnosis, as occurred in our case.

## CASE REPORT

A 59 year old man was admitted through Accident & Emergency with one episode of non-blood stained vomiting and one day history of acute onset lower back and central abdominal pain. The patient reported chronic fatigue and several episodes of lower abdominal pain in the past two years but he had never experienced any gastrointestinal bleeding. Colonoscopy was performed more than a year ago which was normal. His past medical history was remarkable for ischemic heart disease and he had coronary artery bypass grafting (CABG) 11 years ago

On arrival he was haemodynamically stable, his abdomen was soft, non-distended and generally tender. A pulsatile mass was palpable in the midline and around the umbilicus. His routine bloods were normal. Ultrasound scan revealed a 4.4cm infrarenal abdominal aortic aneurysm; however no

definite leak was identified. Computed Tomography (CT) scan of the abdomen and pelvis confirmed the ultrasound findings.

Because the patient was haemodynamically stable and no leak was identified on the scans, it was decided to treat him symptomatically and repair his aneurysm on a later date electively. His pain settled with conservative treatment and he was discharged home after nine days.

A week later he was re-admitted with another episode of non-specific abdominal pain and he was haemodynamically stable. On arrival his blood pressure was 130/74 mmHg and his pulse rate was 76 beats per minute. His abdomen was soft, non-distended and diffusely tender. On the second day, he developed massive haematemesis and went into shock. He was then transferred to the intensive care unit where haemodynamic stabilization was achieved with intravenous fluid replacement, blood and fresh frozen plasma (FFP) transfusions. His blood pressure improved to 90/40 mmHg. An emergency oesophagogastroduodenoscopy (OGD) was then performed, which showed a normal oesophagus with a small amount of dark red blood remaining in the stomach and normal first and second parts of the duodenum with no active bleeding.

After initial resuscitation the patient was transferred to the operating theatre for an emergency laparotomy. A primary aortoduodenal fistula (PADF) was found between the fourth part of the duodenum and the proximal infrarenal aortic aneurysm. Following proximal control of the infrarenal aorta, distal control at both common iliac arteries was obtained. The AAA was repaired using a Gore-Tex® tube graft and direct transverse closure of the duodenum was performed in two layers. An omentopexy was performed between the aorta graft anastomosis and duodenum. The patient received 54 units of blood, 12 units of FFP, 6 units of cryoprecipitate and 12 units of platelets pre and peri operatively. At the end of surgery the patient developed disseminated intravascular coagulation (DIC) and became bradycardic and acidotic, his blood pressure and urine output dropped drastically and he died on the operating table.

## **DISCUSSION**

Primary aortoduodenal fistula is a rare complication of aortic aneurysm and an unusual cause of catastrophic gastrointestinal bleeding [3]. The fistula most commonly erodes into the third part of the duodenum as a consequence of the intimate contact of this enteric segment with the underlying pulsatile aorta. The diagnosis is difficult and

sometimes not reached until laparotomy [3, 5]. Although the typical symptoms of PAEF consist of abdominal pain, gastrointestinal haemorrhage and a pulsatile abdominal mass, but this classic triad of symptoms can only be found in 23% of the patients [11]. The characteristic picture is of a “herald” haemorrhage followed hours, days, or weeks later by catastrophic haemorrhage. The herald bleeding is the result of a small fistula tamponaded by thrombus formation. If the fistula continues to expand or the occluding thrombus is removed massive haemorrhage will occur. Since 70% of patients survive at least 6 hours after the initial bleeding episode and up to 50% survive 24 hours, a herald haemorrhage should be viewed as an opportunity for prompt intervention [1].

An aggressive diagnostic workup and a high index of suspicion are required for successful outcome in cases of PAEF. Endoscopy of the upper gastrointestinal tract should be strongly considered as the first step in diagnosis. The sensitivity of upper gastrointestinal endoscopy in detecting aortoenteric fistula is unknown, but this technique probably identifies less than half of the cases. However, endoscopy may disclose another cause of bleeding. Therefore normal findings or a positive finding of gastritis or ulcers without active bleeding does not rule out aortoduodenal fistula. Careful inspection of the distal duodenum should be performed in the setting of unexplained torrential bleeding [12]. Computed tomography with contrast is the most suitable diagnostic test [13]. The CT may show an abnormal communication between the aorta and the bowel or may disclose loss of continuity of the aneurysmal wall and air bubbles in the aneurysm wall that are pathognomonic for the existence of a fistula [14,15]. Percutaneous angiography may be considered but is rarely of value since most patients are critically ill when the decision for angiography is made [16].

Emergency exploratory laparotomy should be performed as soon as the diagnosis is concluded clinically. Mortality is 100% without surgical intervention [17].

In our case two main points needed to be emphasized (1) the initial decision to defer vascular surgery on an elective basis at the time of aortic aneurysm identification as no leak was identified and patient was haemodynamically stable, (2) PADF was in the fourth portion of the duodenum which is very rare (6%) [18] so that on OGD it was not easily visible. However, it could be argued that even in the best case scenario of early detection of PADF the final outcome would not have been different, given the high mortality in most of

the series [3,4,5, 19].

## CONCLUSION

The diagnosis of PAEF/PADF is difficult and sometimes not made until exploratory laparotomy. A delay in its identification, which is rather common, may partly explain the high mortality. The diagnosis should not only be considered in any patient known to have an abdominal aortic aneurysm but also in a patient who presents with lower abdominal pain associated with a midline mass and exhibits upper gastrointestinal bleeding when no other source of bleeding is seen on OGD.

## LEARNING POINTS

- The diagnosis of primary aortoenteric fistula should be considered in any patient known to have abdominal aortic aneurysm or lower abdominal pain associated with midline mass and upper gastrointestinal bleeding of unexplained aetiology.
- A “herald bleeding” is an opportunity for prompt intervention
- Endoscopy is the first step in diagnosis. Computed tomography may confirm the diagnosis.
- Emergency exploratory laparotomy should be done as soon as the diagnosis is considered.

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