CASE REPORT

Aortocaval fistula: an unusual complication of ruptured abdominal aortic aneurysm

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SUMMARY

Aortocaval fistula (ACF) is a rare complication of abdominal aortic aneurysm, which erodes into the wall of the inferior vena cava, resulting in the formation of a fistula. ACF presents with various inconsistent symptoms and signs. ACF can be a diagnostic dilemma if not suspected and it is lethal if left untreated.

A 60-year-old man presented with abdominal and lower back pain of sudden onset. Renal and liver functions were impaired, without signs of cardiac failure. CT angiography revealed an abdominal aneurysm rupture into the inferior vena cava. Patient underwent a successful open repair: ACF ligation from within the aneurysmal sac and an aorto-bi-iliac bypass using a Dacron graft. Renal and liver functions improved and the patient was discharged on the eighth postoperative day. A month later, the patient was fit and well with normal liver and renal functions.

Despite progress made in the endovascular treatment of ACF, complications still persist with a reported endoleak rate of 50%. Open repair is still a valid method for ACF repair in patients fit to undergo laparotomy and general anaesthesia. In this case, the patient was fit and along the lack of a suitable stent graft, the vascular team performed an open repair with good results.

Open repair of an ACF is a valid treatment method for patients who are fit enough to undergo laparotomy and general anaesthesia, and avoids complications related to endovascular repair, such as endoleaks.

BACKGROUND

Aortocaval fistula (ACF) is a rare complication of abdominal aortic aneurysm (AAA), which erodes into the wall of the inferior vena cava (IVC), resulting in the formation of a fistula.¹ ACF may also result from penetrating trauma and other rarer causes.^{2,3} The first case of ACF was reported in 1831 by Syme.⁴ In 1955, Cooley reported the first successful surgical repair of ACF, while Wooley reported in 1995 the first aortic exclusion used in the repair of an intraoperatively diagnosed ACF.⁴ The first endovascular treatment of ACF was reported by Beveridge *et al* in 1998, while a hybrid technique, using both endovascular and open surgical treatment, was reported in 2009 by Siempre *et al*.^{4,5}

Since 1998, endovascular treatment with stent grafting is preferred due to shorter operative time, lower blood loss and shorter postoperative recovery time.^{6,7} Indicatively, patients who underwent open surgery had a 12% mortality, while patients who

underwent endovascular aneurysm repair (EVAR) had a significantly lower mortality rate of 3.8%.⁴ However, open surgical ligation of the ACF is still performed today, in order to avoid endoleaks, that according to Orion *et al* may persist in 50% of ACF cases after endovascular repair.^{1,4,5} ACF presents with various inconsistent symptoms and signs, and it can be a diagnostic dilemma if not suspected. ACF is a lethal condition if left untreated.⁶

Here, we present a case of a 60-year-old patient with ACF, which was repaired with an open surgery, instead of the lately more common EVAR.

CASE PRESENTATION

A 60-year-old man presented with abdominal and lower back pain of sudden onset at the emergency department. Medical history included a known infrarenal abdominal aortic aneurysm, but the patient did not follow previous instruction for annual follow-ups.

Physical examination revealed a slightly tender abdomen, a pulsating abdominal mass and a purring abdominal sound.

INVESTIGATIONS

Laboratory reports included an increased serum creatinine, SGOT and SGPT levels of 2.7 mg/dL, 1270 U/L and 1108 U/L, respectively.

An urgent abdominal CT angiography (CTA) was performed and an 11 cm aortic aneurysm (figure 1) and an aortocaval fistula at the level of the aortic bifurcation were diagnosed (figure 2). During the arterial phase of the CTA, a significant flow of contrast material from the aorta to the IVC through the ACF was discovered (figures 3 and 4).

DIFFERENTIAL DIAGNOSIS

Differential diagnosis should include the majority of causes of acute abdomen: acute appendicitis, acute cholecystitis, acute pancreatitis, intestinal obstruction, intestinal perforation and mesenteric ischaemia.

TREATMENT

Under general anaesthesia, a midline laparotomy was performed. Infrarenal aortic clamping and digital compression of the inferior cava controlled haemorrhage from the ACF. Then the ACF was ligated from within the aneurysmal sac (figure 4) and an aorto-bi-iliac bypass was performed using a Dacron graft. The patient was discharged home on the eighth postoperative day with normal levels of

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Figure 1 Abdominal aorta and inferior vena cava during arterial phase of CT angiography; white arrow indicates compressed inferior vena cava.

Figure 2 Aortocaval fistula on CT angiography (sagittal view); dark arrow indicates point of communication between the two vessels.

serum creatinine and transaminases, and close-to-normal renal function.

OUTCOME AND FOLLOW-UP

On follow-up a month later, the patient was fit and well, and had laboratory results within normal ranges. Renal function was normal with a mildly decreased glomerular filtration rate.

DISCUSSION

A primary/spontaneous ACF occurs in 1% of AAA cases and 6% of ruptured AAA cases, being thus responsible for 80% of all ACF cases.⁷ AAAs can induce the necrosis of the aortic wall and produce an inflammatory response, which causes the adherence of the IVC to the aortic aneurysmal sac. The rupture of the vessel wall between the two vessels leads to the development of ACF as has happened in our patient.^{8,9} Traumatic ACF, produced by vascular injury in penetrating trauma, makes up 15% of all ACF cases, while iatrogenic ACF, most commonly caused by exploratory laparotomy and lumbar laminectomy, is responsible for 5% of cases.^{7,8} Other rarer causes include mycotic aneurysms,

Figure 3 Aortocaval fistula on CT angiography (coronal view); dark arrow indicates point of communication between the two vessels.

Figure 4 3D reconstruction of CT angiography; dark arrow indicates approximate point of communication between the two vessels.

Takayasu's arteritis and connective tissue diseases, such as Ehlers-Danlos and Marfan syndromes.^{2,3,10} ACF can present with signs and symptoms of varying gravity and character. Liver

congestion, renal failure and right heart congestion are the result of venous hypertension due to high aortic flow into the low-pressure venous circulation.⁶ In most cases, ACF results in sudden death if left untreated due to significant compromise of the heart function,⁶ something that was not present in our case probably due to the good cardiac status of the patient and the degree of the venous congestion. In our case, the patient presented with typical symptoms of AAA rupture, but liver and renal insufficiency, as well as the purring abdominal sound, led the vascular team to suspect the presence of an ACF. Other symptoms, such as phlegmasia cerulea dolens, have also been rarely reported, but in this case lower extremities were not visibly affected by the venous congestion.

Current ACF treatment is accomplished either with open surgical repair or with EVAR.⁷ Open surgical repair consists of rapid suturing of the fistula from within the aneurysmal sac, while endovascular repair is achieved with placement of an endograft at the site of the ACF.¹⁻⁴ Moreover, a hybrid technique can be used, during which an endovascular stent is first placed in the IVC to reduce the central pressure and then the aneurysm can be repaired using the well-known open surgical procedure, provided that immense and significant blood loss from the IVC through the ACF ostium is now halted.⁵⁻⁷ Endovascular repair is beneficial for frail patients and hostile anatomy. On the contrary, open repair is performed when ACF is a coincidental finding during exploratory laparotomy, or when the aortic anatomy is extremely hostile for endovascular treatment. Moreover, open repair can be preferred against EVAR, when comorbidities affecting the cardiovascular and respiratory system are absent. Especially in younger patients, constant follow-up and reintervention, which are often required after endovascular surgery, can be avoided with open repair. In our case, the vascular team decided to proceed with an open repair as no suitable stent graft was available at the time of admission and the patient was fit to undergo a laparotomy. Open repair of an ACF is a valid treatment method for patients who are fit enough to undergo laparotomy or general anaesthesia, and avoids complications related to endovascular repair, such as endoleaks.

Learning points

- Proper follow-up is essential for patients with history of aortic aneurysm (repaired or not).
- In patients with a history of aortic aneurysm presenting with abdominal and/or back pain, aortocaval fistula (ACF) is a valid differential diagnosis.
- Patients with history of aortic aneurysm and pathology increasing venous pressure are more prone to be diagnosed with an ACF.

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