

# A rare presentation of a rare complication of ruptured AAA ... how cardiology comes into play

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## Summary

Ruptured abdominal aortic aneurysm (rAAA) with an arterio-venous fistula is a rare phenomenon. We report a case where the coincidence of significant coronary stenosis was masking the main problem of an infra-renal aortic aneurysm rupture.

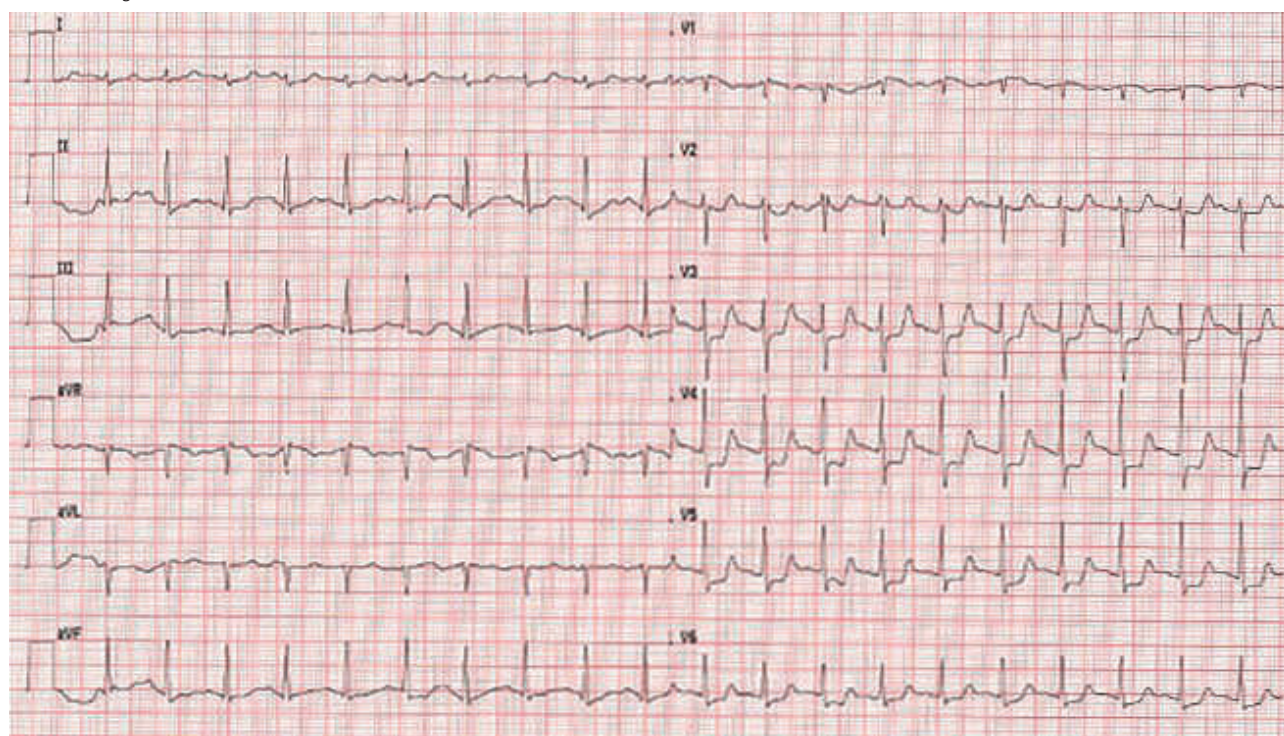
*Key words:* ruptured abdominal aortic aneurysm; aortocaval fistula; acute coronary syndrome; left-sided ivc

## Case report

A 60-year-old man arrived in the emergency room with palpitations, left mandibular pain and presyncope. Some months previously he was diagnosed with chronic lymphatic leukaemia, but otherwise the patient was in good health without any regular medication. He is a smoker (50 pack years) with a family history of coronary heart disease. On arrival the patient presented with a heart rate of 135 bpm and in respiratory distress

**Figure 1**

Electrocardiogram: anterolateral ischemia.



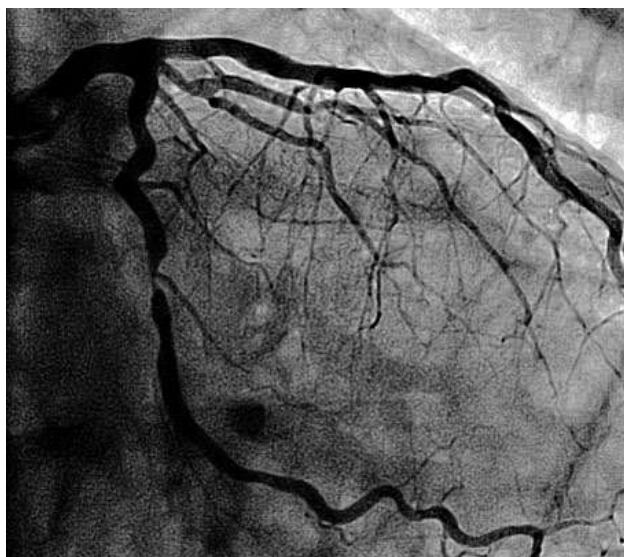
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**Figure 2**

Coronary angiography: significant lesion in the circumflex artery.



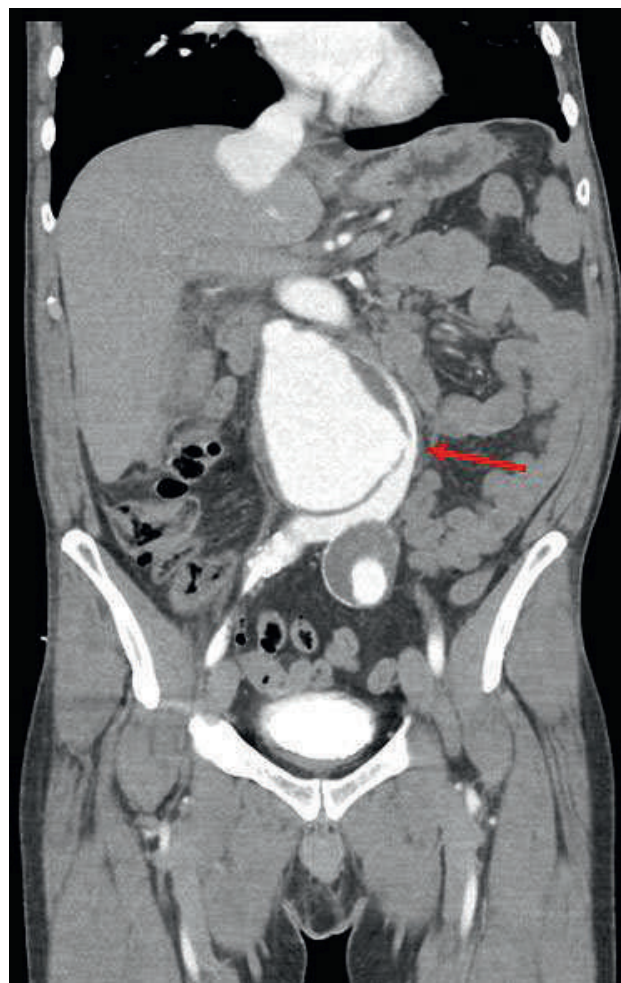
with oxygen saturation of 98%, and blood pressure of 108/53 mm Hg. Distended jugular veins were noted, but neither peripheral oedema nor pathologic heart murmurs were present. The ECG demonstrated sinus tachycardia with significant ST- depressions in V3–6 (fig. 1). Troponine was slightly elevated (0.15 ug/l, reference: <0.1), creatinin kinase was normal (88 U/L) as was brain natriuretic peptide (66 ng/l). Renal function was slightly impaired (119 umol/l), haemoglobin was normal (150 g/l). Trans-thoracic echocardiography (TTE) revealed a hyper-dynamic left ventricle with normal systolic function without wall motion abnormalities. In agreement with the clinical evaluation, TTE demonstrated elevated central venous pressure (distended inferior caval vein) and elevated pulmonary pressure (RV/RA gradient 45 mm Hg).

Due to clinical presentation with signs of the right ventricular overload, pulmonary embolism had been excluded by CT angiogram before we were called to investigate the patient. In light of the global ischemia suggested by the ECG, elevated troponine level and the cardiovascular risk factors (smoking and family history), the decision was made to perform a coronary angiography as acute coronary syndrome was suspected. The patient received aspirine and a loading dose of ticagrelor. The coronary angiography showed a subtotal occlusion of the left circumflex (LCx) (fig. 2) and a moderate stenosis in the right coronary artery. Again, the left ventricular angiogram confirmed a very high cardiac output. While the LCx lesion may explain part of the ECG changes and symptoms, it can not explain the patient's hyper-dynamic shock.

In due course the patient was progressively complaining about lower back pain. Despite the findings in this patients' angiogram the cause of symptoms in this

**Figure 3**

Computed tomography: ruptured abdominal aortic aneurysm with fistula into the left-sided inferior vena cava.



patient remained unclear. Since we had difficulties to guide the catheter from the femoral artery through the aorta, we performed an angiography of the abdominal aorta, which demonstrated a large aneurysm; however image quality was very poor due to the high flow state resulting in significant dilution of the contrast shot – a phenomenon that was not further challenged at that moment. Instead, the patient underwent CT angiography which unveiled a large ruptured infra-renal aortic aneurysm (8.9 cm) communicating with an aberrant left-sided vena cava inferior (fig. 3). The patient was immediately transferred to a tertiary centre to undergo surgery.

A bifurcated Dacron prosthesis was implanted and the fistula involving the inferior caval vein was sutured from inside the arterial wall with Prolene. Intra-operatively the patient obtained 15 blood units, 14 fresh frozen plasma units and 1 thrombocyte concentrate. Initially the postoperative phase was unremarkable. On the 4th postoperative day, the patient developed a paralytic ileus. Due to the small vein lumen

compressed by the large size of the aneurysm the patient got anticoagulation with marcoumar for three months postoperatively. As a consequence of mass blood transfusion, the patient's blood group converted from 0 negative to 0 positive.

The patient was discharged after 10 days, subsequently participated in an outpatient rehabilitation programme at our hospital and the significant stenosis of the LCx was treated by stent implantation 7 weeks after the vascular operation.

## Discussion

Rupture of abdominal aortic aneurysm with spontaneous aortocaval fistula is very rare. It was first described by Syme in 1831 and it took many years until the first successful operation of a fistulating abdominal aortic aneurysm was performed by Cooley in 1954 [1, 2]. Most publications are case reports or reviews, reporting an incidence of 0.22–6% of spontaneous aortocaval fistula in presence of abdominal aortic aneurysm rupture [5–8]. Patients usually are male and 60 to 70 years old [4–5, 7–10].

The arteriovenous short circuit dramatically reduces the peripheral resistance which subsequently leads to hyper-dynamic cardiac shock/failure as well as hypoperfusion of the kidneys with renal insufficiency [3, 5, 7]. Cardiac arrest may occur, in particular in the presence of coronary artery disease. Decision making in these patients can be difficult as symptoms of circulatory and respiratory failure mask the classic symptoms of abdominal aortic aneurysm rupture with features of haemorrhagic shock with lower back pain, abdominal pain, palpitations. Clinical examination may show a pulsating abdominal mass with bruits, an elevated central venous pressure, oedemas, hematuria or oliguria and scrotal oedemas [5–7, 9–10]. Deep palpation and auscultation of the abdomen in all quadrants, inguinal region and the kidneys should be considered in every patient.

Most patients with an abdominal aortic aneurysm and arterio-venous fistulas rupture into the inferior vena cava whereas rupture into an iliacal vein is rare [3–5, 9–10]. In most reports the aortic aneurysm ruptured into a normal inferior vena cava and there is not much data about fistulas with aberrant caval veins.

Besides atherosclerosis, which is the aetiology in most cases, trauma, interventions, infectious diseases like syphilis connective tissue disorders (Marfan- and Ehlers-Danlos-syndrome), Takayasu arteriitis or neoplasms may cause fistulating aneurysms [3–6, 10].

Rapid diagnostic assessment, computed tomography or ultrasound [4, 6], and surgical intervention are crucial [3]. Reported mortality rates are in the range of 16–66% [4–5, 9] and significantly higher than with rupture into the retroperitoneal cavity [3–4, 7].

In our case the initial right heart volume overload

with sinustachycardia suggested pulmonary embolism first, which was ruled out immediately by a CT scan. Having excluded pulmonary embolism, the ST-segment depressions in antero-lateral leads on ECG, the elevated troponine, the pain in the left mandibula and the known cardiovascular risk profile suggested acute myocardial infarction with cardiogenic shock. The classic symptom of rAAA with low back pain, which would have been important for early diagnosis, developed later during coronary angiography. The initial clinical findings were discrete; a pulsating abdominal mass or absent pulses were not noted during initial presentation. This may be related to the reduced peripheral resistance caused by the fistula into the vena cava. Presence of an aortic aneurysm was first suspected during the coronary angiography and confirmed by aortography even though significant coronary lesions were present correspondent to the ECG. The association between coronary heart disease and abdominal aortic aneurysm is well known. About 43% of patients with abdominal aortic aneurysm have co-existing coronary heart disease [11], which impacts perioperative risk and mortality [3, 8–9].

One could argue that in context of a normal echo without wall motion abnormalities, coronary angiogram shouldn't have been prioritised in this situation. An abdominal ultrasound or CT would have been sufficient to solve the diagnostic problem if there would not have been the misleading signs of acute coronary syndrome. Haemoglobin was high normal, significant bleeding despite ruptured AAA was not present as the aneurysm fistulated into the venous system and consequently revealed an atypical rupture of abdominal aneurysm.

With the imaging studies performed, the contrast medium load was 314 ml, which is a concern in the setting of impaired renal function. As the patient presented life threatening conditions, further diagnostics including contrast studies was mandatory. The CT-Angio of the abdominal aorta was required for the surgical planning irrespective of ultrasound findings. Given the ECG-changes and initial clinical presentation of the patient, the vascular surgeon felt comfortable to be familiar with the coronary anatomy for his high-risk procedure. Of course, one should argue that dual anti-platelet therapy should not be started in unclear situations and postponed until coronary anatomy is clarified. Dual anti-platelet therapy complicates surgical procedures including coronary artery bypass grafting. However, all present algorithms of acute coronary syndrome include early dual anti-platelet therapy.

The key points of this case report are that even in presence of a ruptured abdominal aortic aneurysm this entity may be masked by unusual anatomical conditions and clinical presentation. In our patient, the rupture occurred into an aberrant inferior caval vein by a fistula changing the clinical presentation significantly.

Moreover, coronary heart disease misled us. Nonetheless the outcome of this patient was good due to the fact that a recirculation of blood into the venous system occurred.

In this particular case, the high cardiac output, which cannot be explained by acute coronary syndrome, was present but not taken into the initial considerations. This case emphasises the importance of a thorough clinical examination before starting diagnostic machinery. This may be even more important in stressful situations with a shocked patient with an unusual clinical presentation.

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