

Bursting heart syndrome: A rare case of aortocaval fistula repair

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Abstract

In this case report we will discuss the case of a 47-year-old woman who presented with abdominal pain, nausea, oliguria and right heart failure. A Computed Tomography (CT) aortogram revealed a fistulous abdominal aortic aneurysm.

The objective of this study is to discuss the haemodynamic changes regarding aortocaval fistula and consider various management options.

A literature search was undertaken on PubMed using appropriate search terms. Case series and reviews reporting presentation, diagnosis and operative management of aortocaval fistula were selected and discussed.

We reached a conclusion that early identification improves surgical planning and reduces mortality. Major abdominal arteriovenous fistula repair appears to be a safer and more successful therapy with positive short and midterm outcomes. Aortocaval fistula care requires a more extensive patient series, so even better conclusions can be drawn.

Keywords: Inferior vena cava; Aortocaval fistula; Abdominal aortic aneurysm; Open aneurysm repair; EVAR; Hybrid abdominal aortic aneurysm repair.

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Introduction

Aortocaval fistula (ACF) is an abnormal connection between the aorta and inferior vena cava. This life-threatening condition has less than 1% incidence; however, it is 2-3% more common in ruptured abdominal aortic aneurysms.¹ Aortocaval fistula can be primary, caused by an aortoiliac aneurysm eroding into the venous system, or secondary due to iatrogenic or traumatic conditions.² This rare condition can cause various symptoms, and diagnosis is often delayed or overlooked. Treatment of this anomaly is still debated however endovascular stent-graft repair seems to offer an alternative to open surgery for this life-

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threatening condition. We will discuss the case of a 47-year-old woman who presented with abdominal pain, nausea, oliguria, and right heart failure. A CT aortogram revealed a fistulous abdominal aortic aneurysm. We will focus on the case's haemodynamic changes, technical issues, and complications.

Case Report

A 47-year-old diabetic, hypertensive and morbidly obese woman presented at emergency department of Shifa International Hospital Islamabad on 26th May 2022 with complains of abdominal pain, nausea, and oliguria for ten days. The patient had generalized swelling all over her body with marked swelling of the lower limbs. Her blood pressure was 80/40mmHg and her respiratory rate was 20/min. Her heart rate was 64/min and a Central Venous Pressure (CVP) of 29cm. She had generalized oedema with marked swelling in the legs. A CT scan revealed a large infra-renal abdominal aortic aneurysm extending into bilateral common iliac arteries on the right, measuring approximately 8.3x9.7x11.5 cm. Early filling of inferior vena cava suggested aortocaval fistula (figure 1). Hepatomegaly was also present.

The laboratory values revealed that Creatinine was

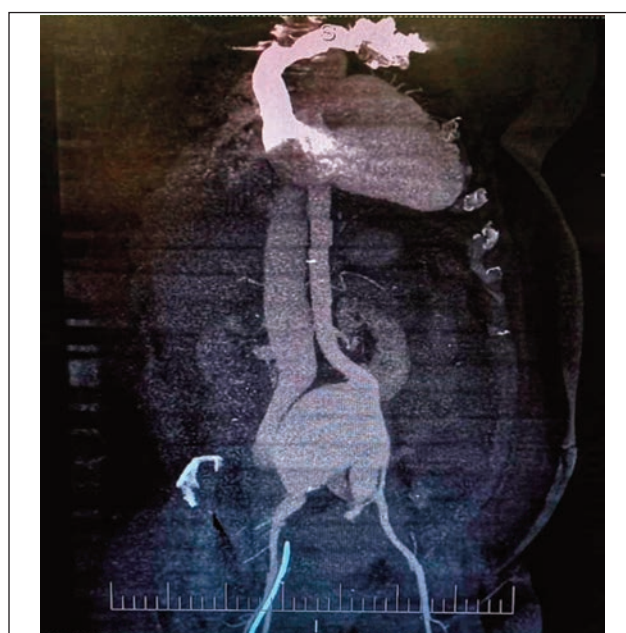


Figure-1: CT aortogram showing Large infrarenal abdominal aortic aneurysm extending into bilateral common iliac arteries.



Figure-1: Repair aortocaval fistula with Dacron graft.

4.9 mg/dl (normal range 0.7-1.2 mg/dl) and 24-hour urine output was found to be 150ml. The liver functions tests showed that bilirubin was 1.1 mg/dl (normal range 0-1mg/dl), AST was 1219 (normal range 10-40IU/L) and ALT was 1466 (normal range 10-40IU/L). An echocardiography was done in the emergency room and it showed that there was dilatation of the right heart, severe tricuspid and pulmonary regurgitation and an ejection fraction of 55%.

A plan of open surgical repair was made. Anaesthesia team labelled the patient as ASA grade 4. A midline laparotomy was done. Aorta and the aneurysm were identified, dissected and cross-clamps were applied proximally and distally. Once the control was established, the sac was opened. There was a torrential bleed from the sac and the fistula was identified. The venous bleeding from the sac was controlled by applying direct pressure. The fistula was oversewn from within the aneurysmal sac with Prolene 3-0. The aneurysm was repaired with an 8*16cm aorto-biiliac dacron graft (figure 2). The patient was returned to ICU after operation for further management. The patient needed inotropic support and was on a ventilator for four days. Three cycles of dialysis were done in the post-operative period.

Post operatively the haemodynamic status of the patient started improving dramatically. On 4th post-operative day the creatinine had decreased to 2.6mg/dl and urine output had increased to 25ml/hour. The creatinine further reduced to 1.54mg and urine output was more than 30ml/hour on the 9th post-operative day before the patient was discharged. The liver enzymes showed a substantial

improvement as well. On the 2nd post-operative day, AST and ALT had come down to 145 and 260 respectively normal range 10-40IU/L. The cardiac status of the patient showed a remarkable improvement. CVP of the patient decreased from 29mm Hg to 14mm Hg normal range 8-12mmHg on the 5th post-operative day. An Echo done before discharging the patient showed that there was dilatation of right atrium only with no pulmonary or tricuspid regurgitation present.

The patient was discharged on the 10th post-operative day. She had two follow ups till date. She is doing fine and the wound has healed pretty well.

The histopathology of the aneurysmal sac wall showed vessel wall calcification with fibrosis and scattered lymphoid aggregates indicating that the cause was atherosclerosis.

Discussion

Primary ACF, caused by an aortoiliac aneurysm eroding into the vena cava causes most of the Aortocaval fistulas³ with atherosclerosis being the most common cause. Occasionally, rupture of syphilitic or mycotic aneurysms⁴ as well as the ones caused by Marfan's Syndrome, Ehlers-Danlos Syndrome, and Takayasu's Arteritis have been reported to cause ACF.⁵ Fistulas caused by aneurysms are usually aortocaval.

Abdominal Aortic Aneurysm (AAA) with ACF has many ambiguous symptoms. Pulsatile abdominal mass, low-back pain, and abdominal bruit are commonly present. Other symptoms include severe dyspnoea, scrotal or lower extremity oedema, ascites, hepatomegaly, liver congestion from venous hypertension, haematuria, oliguria, renal failure, and low median arterial pressure. These symptoms are caused by low peripheral resistance. Our patient developed abdominal pain, right-sided heart failure, bilateral lower limb oedema, and anuria. High mortality is due to high output cardiac failure. These patients also have pulmonary embolism. Contrast-enhanced aortography usually confirms the diagnosis.

Once diagnosed, immediate surgery is required. A misdiagnosis can be fatal for an elderly patient due to excessive haemodynamic stress. Without fixing ACF, improving cardiac or renal function is futile. Any patient with cardiac and renal compromise is an indication for surgery.⁶ Open or endovascular repair is possible. Our patient was treated with open surgery, which has high morbidity and mortality. Minimal aortic aneurysm handling is advised during surgery to avoid intraluminal debris embolization. Excessive bleeding is another problem. To control bleeding, supra-coeliac aortic clamping may be

needed. Because of the risk of renal ischaemia, it is not recommended unless there is uncontrolled bleeding. After opening the aorta, venous bleeding must be controlled. Digital pressure or occlusive balloons are used to do this. We used digital pressure to control bleeding while repairing IVC. Two Foley or Fogarty balloon catheters can be inserted through the fistula or femoral vein to occlude the IVC lumen. Effective treatment reduces decompensation and restores renal function. Even in semi-elective cases, the death rate approaches 30%, especially in cardiovascular decompensation.⁷

Endovascular repair has gained popularity since successful trial treatment on eight sheep.⁸ It is complicated by type 2 endoleak caused by retrograde sac filling. Antoniou's 2009 research showed 96% technical success (22/23) with no peri-operative or 30-day mortality over a 9-month follow-up. 23% (5/23) of patients had type II endoleaks.⁹ This condition resolved itself or required minimal percutaneous treatment. Lau et al. (2001) found that occlusion of ACF upon stent graft release causes a sudden increase in Systemic Vascular Resistance. Hypertension refractory to medical treatment may require invasive monitoring. This shift is not seen after open surgery due to aortic cross-clamping and intraoperative haemorrhage. Endovascular procedures have less blood loss than open surgery, reducing mortality.¹⁰ Unlike a AAA rupture or confined rupture, spontaneous ACF may not always require emergency care, and time may be available to procure an endovascular stent-graft.

In 2009, Siepe et al. reported a hybrid approach. The patient was unsuitable for endovascular repair because of haemodynamic instability. In order to treat the fistula, Siepe et al. decided to insert a sizable covered aortic stent into the IVC. The central pressures were successfully decreased from 33 to 19 mm Hg, arterial blood pressure was stabilized, and inotropic support could be withdrawn. As a result of the patient's stabilization, the AAA was operated on using routine open surgery without running the danger of severe blood loss due to the significant IVC defect.¹¹

Conclusion

Aortocaval Fistula is not a common condition causing misdiagnosis. Identification before surgery reduces mortality and improves surgical planning. Endovascular

exclusion of AAA or IVC can improve haemodynamic status. Repairing major abdominal arteriovenous fistulae is a safer and more successful therapeutic choice with positive short and midterm outcomes. More extensive patient series are needed to draw firm conclusions about Aortocaval Fistula care. Reviewing previous cases is another option. This would allow for better judgments and management standards.

Consent: Verbal consent was obtained from the patient for publishing her case.

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