Acquired Arteriovenous Fistula of the Right Common Iliac Artery and Left Common Iliac Vein and Bilateral Lower Extremity Deep Venous Thrombosis in a Woman Presenting as High Output Heart Failure

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ABSTRACT

Acquired intraabdominal arteriovenous fistula (AVF) is a rare disorder where the communication most commonly occurs between the abdominal aorta and inferior vena cava. Ilioiliac AVF has been reported previously, but is exceedingly rare. We present a case of acquired arteriovenous fistula of the right common iliac artery and left common iliac vein with extensive collateralization (ilioiliac AVF) in a 36-year-old female who presented with symptoms of high output congestive heart failure 18 years after sustaining an abdominal gunshot wound.

Key Words: high output heart failure, arterio-venous fistula, deep venous thrombosis, ilio-iliac, aortocaval fistula, gunshot wound, trauma

Introduction

Arteriovenous fistula (AVF) involving major vessels is a rare clinical entity that may lead to severe hemodynamic disturbances.[1] The AVFs are most commonly aorto-caval fistula, followed by ilio-iliac[2] and aorto-iliac. The aetiology, clinical features, pathophysiology, principles of management and postoperative care for these fistulas are similar.[3]

Most patients either die from exsanguination; however, if they survive the initial acute period, the natural course of the fistula is to enlarge over time and only rarely close spontaneously. This results in a myriad of severe hemodynamic disturbances and progressive clinical deterioration.[2]

We report the case of our patient who presented with high output heart failure due to a traumatic fistula connecting the right common iliac artery and left common iliac vein with associated bilateral lower extremity deep venous thrombosis. The clinical presentation, compensatory hemodynamic and structural changes, and ideal management are discussed.

Case Presentation

A 36-year-old female presented to the emergency room with complaints of shortness of breath and abdominal enlargement. Eighteen years prior to admission, she sustained a gunshot wound through the abdomen and underwent surgical exploration. Her recovery was uneventful. Over the past 5 years prior to consult, she had been experiencing progressive exertional dyspnea, two-pillow orthopnea, paroxysmal nocturnal dyspnea associated with abdominal enlargement and intermittent edema of both lower extremities. In spite of this, no consult was done and no medications were taken. In the interim, her functional capacity dropped experiencing dyspnea with minimal activity and the lower extremity edema increased with the left lower extremity more swollen than the right. Two weeks prior to admission, she experienced dyspnea at rest prompting consult in our institution.

Physical Examination

The patient came to the emergency room in mild cardiorespiratory distress, with stable vital signs, blood pressure of 130/40 mmHg, heart rate 104 bpm. On physical examination, there was note of neck vein engorgement and crackles on bilateral lung fields. Cardiac examination revealed a displaced apex beat at the level of the 6th intercostal space anterior axillary line with an associated right and left ventricular heave. There was also note of a

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grade 5/6 systolic murmur at the level of the 4th intercostal space left parasternal border that increased with inspiration. The abdomen was globular with positive fluid wave test. There was a scar on the right lower quadrant of the abdomen representing the alleged entry point of the bullet, with a surgical scar on the right hemi-abdomen (Figure 1). A continuous, harsh bruit was heard over the entire abdomen. There was also grade 3 bipedal, pitting, edema more prominent on the left, reaching the level of the thigh. There was also an ulceration on the left lower extremity (Figure 2). The pulses of all extremities were full and equal.

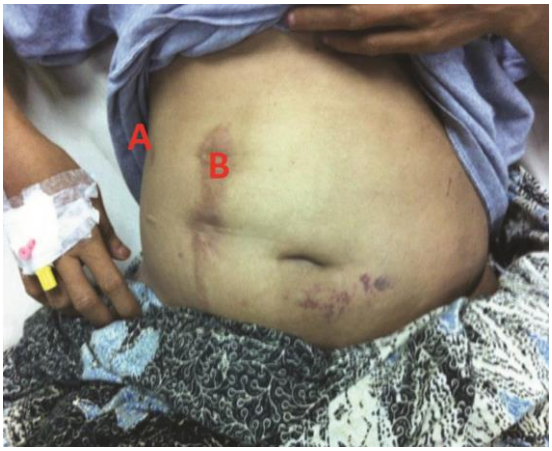

Figure 1. Globular abdomen with 2-cm right abdominal scar representing the alleged entry point of the bullet (A), with a surgical scar on the right hemi-abdomen (B).

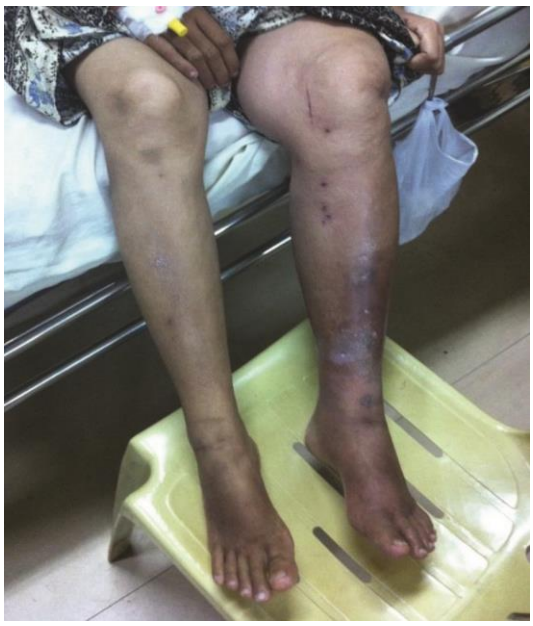

Figure 2. Pitting bipedal edema more prominent on the left with an associated ulceration on the left lower extremity (Picture taken after diuresis).

Laboratory and Diagnostic Studies

Electrocardiogram was done upon admission which showed regular sinus rhythm, right axis deviation, right atrial abnormality, bi-ventricular hypertrophy with strain pattern. Chest x-ray showed marked multichambered cardiomegaly with pulmonary congestive changes (Figure 3). Echocardiogram was subsequently done which revealed concentric left ventricular hypertrophy with good wall motion and contractility and preserved overall systolic function, dilated right atrium and right ventricle (RV) with adequate RV contractility, moderate tricuspid regurgitation, moderate–severe pulmonary hypertension, with a markedly dilated and plethoric inferior vena cava. Holoabdominal ultrasound was initially as an aortocaval fistula formation with resultant marked dilatation of the inferior vena cava and its tributaries.

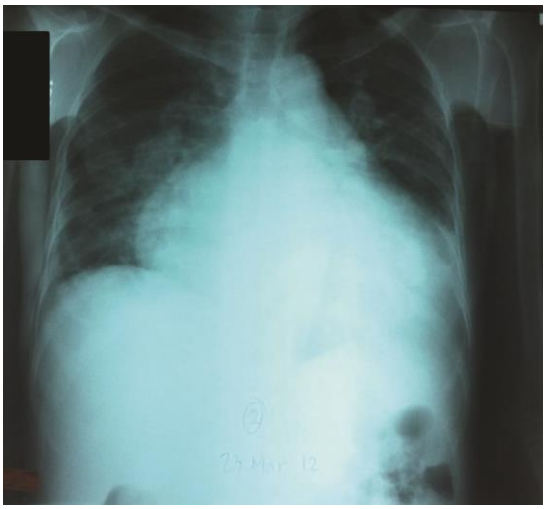

Figure 3. Chest x-ray showing cardiomegaly (Cardiothoracic ratio of 0.74) with evidence of pulmonary congestive changes.

CT aortogram was performed, the findings of which were compatible with an arteriovienous fistula of the right common iliac artery and left common iliac vein with extensive collateralization (Figures 4–10). There was also aneurysmal dilatation of the right common iliac artery (measuring 1.9 x 1.7 cm by its maximum dimensions) and severe fusiform ectasia of the left common iliac vein (maximal diameters of 8.7 x 6.6 cm). This was associated with severe inferior vena cava ectasia more prominent at the infrarenal level (maximum diameters of 8.7 x 6.6 cm) (Figures 4, 6, and 10). Also seen were dilated and serpiginous vessels at the right parametrial region arising from the right internal iliac artery (Figures 8 and 10). Also noted were tortuous dilated vessels along the left pudendal region/ labia majora and left medial thigh. There was a slightly-dilated vein arising from the left renal vein, likely the left ovarian vein, coursing at the left paravertebral

region, extending to the midline, terminating to a tuft of serpiginous vessels at the left iliac fossa. There was also marked multichambered cardiomegaly with congestive changes and dilated pulmonary artery and veins (Figures 9 and 10). A metallic structure was noted at the left sacroiliac joint suggesting a post-traumatic etiology probably a bullet slug (Figure 7).

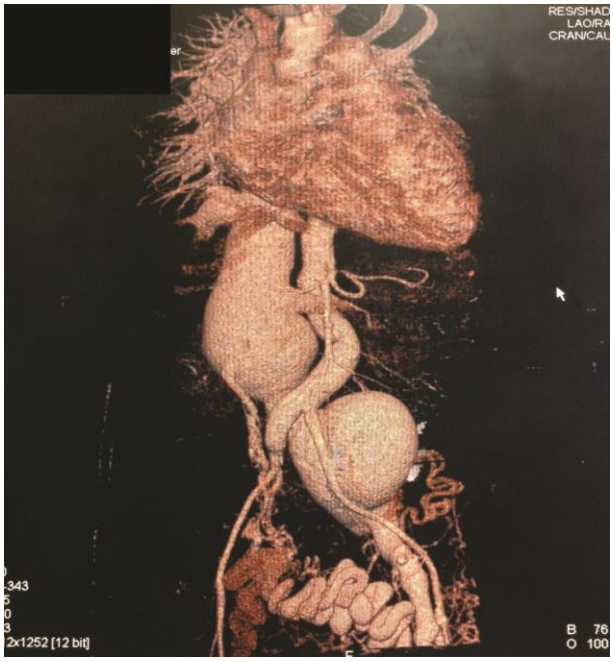

Figure 4. 3D reconstruction showing vascular anatomy. The inferior vena cava is dilated and tortuous. Note how the abdominal aorta crossess over the path of the inferior vena cava producing severe dilatation in its proximal and distal segments

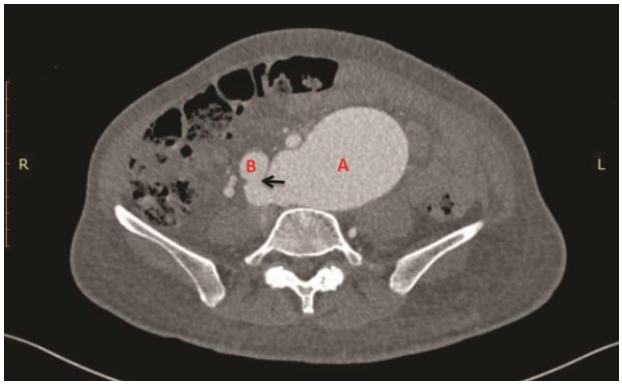

Figure 5. Axial contrast enhanced CT: A fistulous connection (black arrow) of the dilated left common iliac vein (A) to the aneurysmally dilated right common iliac artery is noted (B). The entire right common iliac artery (B) is dilated and lobulated in its contour with maximum dimensions of 1.9 x 1.7cm (W x AP). Fusiform dilatation of the left common iliac vein (A) with maximal diameters of 8.7 x 6.6cm (W x AP).

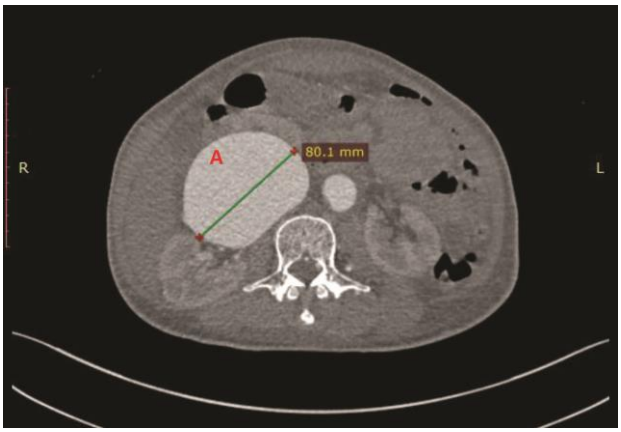

Figure 6. Axial contrast enhanced CT scan showing the aneurysmally dilated and tortuous inferior vena cava (A) with focal and fusiform ectasia involving 11.4 cm of its length at the infrarenal level; maximal diameters of 8 x 6.6 cm (W x AP).

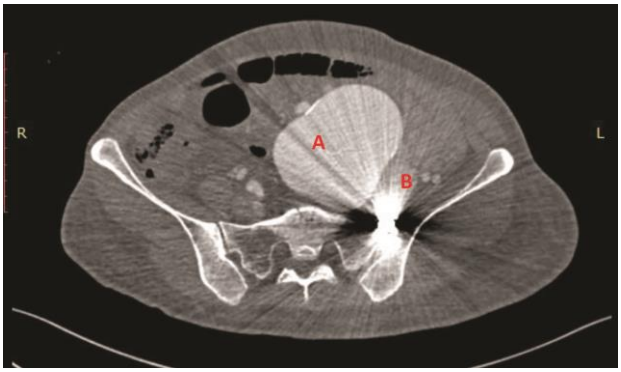

Figure 7. A metallic artifact (bullet slug) is seen at the area of the left sacroiliac joint (A). Fusiform dilatation involving 10.4cm length of the left common iliac vein with maximal diameters of 8.7 x 6.6cm (W x AP) (B)

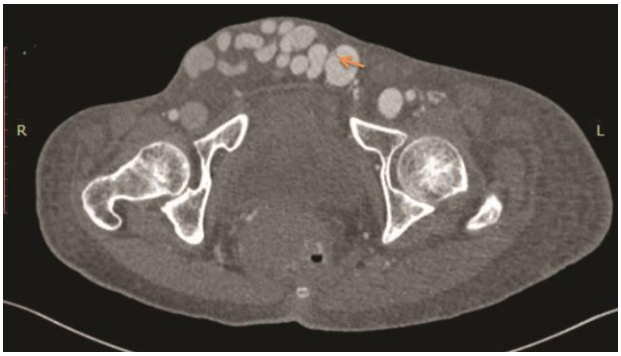

Figure 8. Dilated and serpiginous vessels at the right parametrial region arising from the right internal iliac artery signifying extensive collateralization (colored arrow).

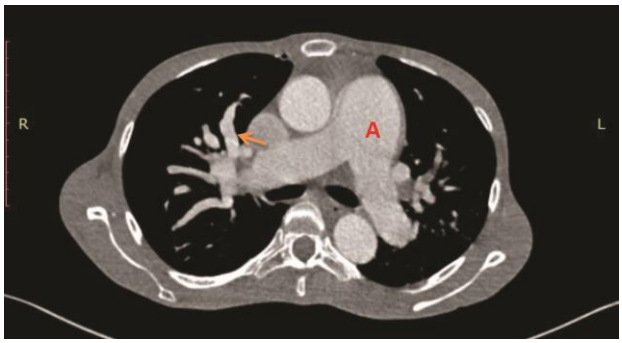

Figure 9. The main trunk of the pulmonary arteries (A) (maximal diameter=3.8cm) as well as its terminal branches are dilated (maximal diameter of the right and left main pulmonary arteries=2.8cm and 2.6cm). There is slight abrupt tapering of their terminal peripheral branches (colored arrow).

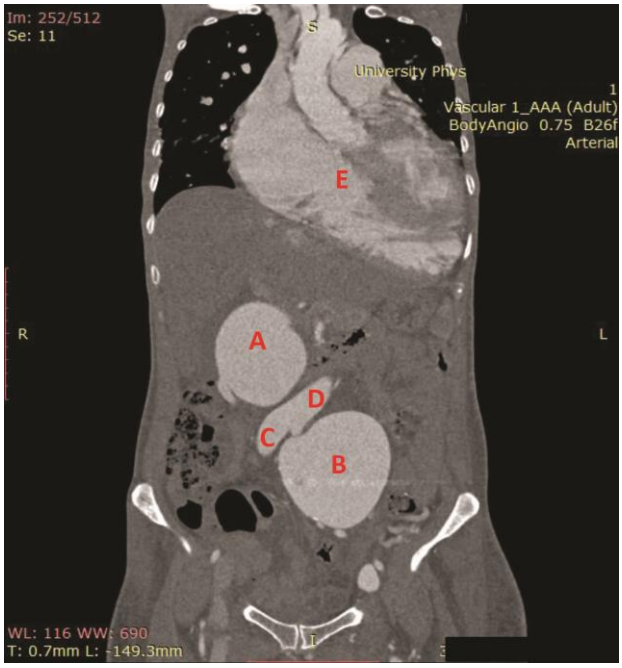

Figure 10. Showing the aneurysmally dilated and tortuous inferior vena cava (A) with focal and fusiform ectasia involving 11.4 cm of its length at the infrarenal level. The dilated left common iliac vein (B) and the aneurysmally dilated right common iliac artery (C) is noted. The abdominal aorta (D) is normal in size, but with slight tortuosity in its infrarenal portions. Marked enlargement of the heart is also seen in all of its chambers (E) especially the atria (right and left) and thickening of the left ventricular myocardium.

Lower extremity venous duplex scan showed right lower extremity deep venous thrombosis (DVT) of the common and superficial femoral veins, acute partial occlusion, with deep venous insufficiency and perforator incompetence; and left lower extremity DVT of the Iliac vein. The final diagnosis was congestive heart failure functional class III from high output heart failure secondary to ilioiliac fistula (right common iliac artery and left common iliac vein), right common iliac artery aneurysm and bilateral DVT. Other blood tests done included complete blood count, blood chemistry, electrolytes, and bleeding parameters which were unremarkable.

Outcome and Management

Surgical repair was strongly advised; however, the patient refused surgery. The patient was managed with optimal heart failure medical therapy: intravenous and oral diuretics, ACE-inhibitors and beta blockers which remarkably reduced the edema and improved her functional capacity after less than a week of administration. She was subsequently discharged with maximal heart failure medications and oral anticoagulation.

Discussion

Epidemiology

Acquired intraabdominal arteriovenous fistulas (AVFs) are uncommon disorders where the communication most commonly occurs between the abdominal aorta and inferior vena cava. Ilioiliac AVF has been reported previously, but is exceedingly rare.[3] Posttraumatic arteriovenous fistulas account for 10% of all cases of traumatic vascular injury and result primarily from gunshot and stab wounds. Approximately 80% to 90% occur as a result of rupture or erosion of an abdominal aortic aneurysm into the inferior vena cava, and 10% to 20% are usually the result of penetrating or iatrogenic trauma.[4,5] The AVFs are most commonly aorto-caval fistula, followed by ilio-iliac[2] and aorto-iliac. However, the etiology, clinical features, pathophysiology, principles of management, and postoperative care for these fistulas are similar.[6]

Clinical Presentation

Two modes of presentation for patients with large traumatic arteriovenous fistulas were identified from previous studies.[4] Most commonly, patients die at the outset from exsanguination. This is associated with an increased mortality ranging from 40% to 45% and is usually the result of the combination of associated injuries, difficulties obtaining exposure, intraoperative exsanguination, hypothermia, and coagulopathy from prolonged surgery.[4,6,7]

The second, less common mode of presentation is delayed and occurs usually weeks, months, or years after the initial injury,[4] in our patient who came in with florid high-output congestive heart failure (CHF) eighteen years after sustaining an abdominal gunshot wound injury.

The timing, progression, and the compensatory changes depend on the size of the fistula, the diameter of the component artery and vein, the proximity to the heart, and the age of the patient. Those who will likely have a more delayed clinical presentation are young and healthy patients with no comorbid conditions because they have very little intrinsic cardiac disease. Furthermore, they are more able to adapt to the hemodynamic changes that result from the fistula.[4,8]

Ilioiliac and aortocaval fistulas rarely close spontaneously. The natural course is to enlarge and develop a continuous bruit or thrill and volume overload.

These AVFs have elusive clinical presentations and the classic triad of abdominal/back pain, pulsatile mass, and abdominal machinery bruit is present only in 20–50% of cases.[9] Other physical findings associated with an ilioiliac fistula include jugular venous distension, pulmonary edema, ascites, pulsating varicose veins, and lower extremity edema.[10,11] Venous hypertension and obstruction distal to the fistula, along with compression of the great veins secondary to the aneurysm, can lead to the clinical manifestations of bilateral swollen cyanosed legs, hematuria or hematochezia, venous stasis, and increased risk of developing venous thrombosis.[11]

Hemodynamics and Pathophysiology

Aneurysmal dilation of the right common iliac artery and fusiform ectasia of the left common iliac vein is a result of the increase in blood-flow velocity and shear stress in both vessels. Both the artery and vein proximal to the AV fistula undergo structural changes in response to the altered hemodynamic stress. Histologically, this is characterized as an increase in collagen and elastin elements in the media of the artery and the vein and more vaso vasorum in the venous adventitia. However, these changes eventually lead to degenerative changes with atrophy of smooth muscle fibers, fragmentation, and a reduction in the number of elastic elements of the arterial wall. The formation of an atherosclerotic plaque is the ultimate consequence of these processes and structural changes may be irreversible if the fistula persists for prolonged periods.[12]

Furthermore, the atherosclerosis and calcification evident on the CT scan is of considerable interest in view of the potential contribution of hemodynamic flow disturbances to the development of atherosclerotic aneurysms. Whether the calcification in the aorta observed was the end result of prolonged hemodynamic injury or merely evidence of heterotopic or dystrophic calcification of the intramural or periadventitial hematoma as a result of the initial injury is unclear.[4]

The physiological changes associated with these fistulas include an increase in pulse rate, a decrease in blood pressure, and an increase in cardiac silhouette. The cardiomegaly was accompanied by a corresponding increase in the diameter of the aorta and vena cava proximal to the fistula. Several factors have been implicated in the dilatation of the aorta and vena cava proximal to the fistula. The most obvious physiologic change concomitant with the formation of an AV fistula is the several-fold increase in blood flow, especially during diastole.[12,13] Other etiologic factors implicated in the aortic dilatation associated with an ACF include vibratory forces and impairment of the nutrition of the arterial wall as a result of changes in the number and diameter of the vaso vasorum. The role cellular signals, metalloproteinases, nitric oxide, and other unrecognized substances in the structural changes that cause the artery to dilate and elongate have been identified by previous studies. Observational reports recognized reversal of the dilatation and elongation of the arterial component after surgical correction of the fistula in the symptomatic patient.[13]

Cardiac decompensation occurs in about 75% of patients. Shunting of blood from a high-resistance arterial circuit to a low-resistance venous system leads to an increased cardiac preload, together with decreased afterload, and ultimately results in a compensatory increase in cardiac output and heart failure.[14] As opposed to the reversal of dilation and elongation of the arterial component after surgical repair, the cardiac silhouette may remain enlarged up to 2 years postoperatively.[4,13,14] This can be explained by the sustained volume overload caused by an AV fistula which produces cardiac myocyte hypertrophy, biventricular dilation, and ultimately, ventricular decompensation. Prolonged hemodynamic stress results in alterations in the remodeling of the ventricular wall, which leads to irreversible cardiomegaly.[4,15] The mechanism responsible for this change in myocardial function in patients with high output cardiac failure associated with AV fistulas may be mediated via the renin–angiotensin mechanism.[14] This can be potentially managed medically by conventional medications recommended for heart failure. The administration of an angiotensin-converting enzyme inhibitor may improve myocardial function but their utility for decreasing cardiomegaly in symptomatic patients or whether these agents could be used to prevent decompensation in asymptomatic patients remains unclear because of the lack of studies for specific patients with this condition.[4,14]

Workup

The standard preoperative imaging technique used in patients with abnormalities of the abdominal aorta and inferior vena cava has traditionally been cut film or digital subtraction catheter angiography.[3]

Multi-slice helical CT scanning has largely replaced conventional angiography as the gold standard for the imaging of aortic pathology however this cannot always be performed with patients with a history of allergic reactions to contrast or increased risk of contrast nephropathy.

Alternatives to CT angiography include magnetic resonance angiography and digital subtraction angiography with carbon dioxide as a contrast agent.[1] Color Doppler ultrasound has demonstrated the AV fistulas as areas of high velocity turbulent flow with aliasing of color signal and also shows any associated thrombus at the aneurysm or fistula. Detection of ilioiliac fistulas may however be difficult as these tortuous and aneurysmal vessels lie deep within in the pelvis and obscured by overlying bowel gas. [3]

Management

The treatment of traumatic or secondary ACF is presently surgical closure of the fistula. As can be seen on the CT scan of these patients, the vena cava is extremely enlarged. Following the closure of the fistula, a prosthetic graft is used to reconstruct the aortoiliac aneurysm. Careful assessment of graft size is necessary, especially because the dimensions of the aorta can be expected to decrease to normal once the fistula has closed. Those with significant high-risk comorbid factors may be considered for treatment by endovascular approach. However, the long-term results of endograft exclusion of aortocaval fistula remain unclear.[16]

Summary and Conclusion

The case described here is an example of progressively deteriorating hyperdynamic heart failure due to the chronic, sustained volume overload caused by a traumatic AVF, specifically connecting right common iliac artery and left common iliac vein seen by helical CT imaging. It represents the clinical extreme of these cases. A thorough history and a physical examination are still indispensible tools that aid the physician in diagnosing such an uncommon condition. In conclusion, it is prudent to include AVFs (ilioiliac, aortocaval, aortoiliac fistulas) as part of the differentials of patients with a history of penetrating abdominal injury or surgery presenting with signs and symptoms of progressive cardiac decompensation, abdominal bruits, and other signs of high output heart failure. It is also a testament to how effectively the body can adapt and compensate to unusual stresses and still function to a degree, before eventually succumbing to the chronic complications and hemodynamic strain of the AVF.

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