Traumatic Peri-Renal AV Fistula Presenting with Pulmonary Hypertension and Right-Side Heart Failure

Chun-Cheng Wang,1 Yu-Shien Ko,1,3 Tsu-Shiu Hsu,1 Kee-Min Yeow2 and Chi-Tai Kuo1

We describe a 47-year-old male patient who presented with symptomatic pulmonary hypertension in association with traumatic peri-renal arteriovenous (AV) fistulae. He developed symptoms and clinical evidence of pulmonary hypertension, and high-output right heart failure long after a left flank stabbing injury 16 years ago. Numerous AV fistulae were illustrated around the atrophic left kidney by spiral computed tomography. Although acquired systemic AV fistula has been implied in the substrates of pulmonary hypertension, association with traumatic AV fistula in the peri-renal region has not been reported previously.

Key Words: Traumatic AV fistula • Pulmonary Hypertension • Right-side heart failure

CASE REPORT

A 47-year-old male was admitted to Chang-Gung Memorial Hospital for slowly progressive exertional dyspnea, orthopnea, bilateral leg edema, ascites and jaundice over 3 years. He had a history of laparotomy for a left flank stabbing injury 16 years before. Another operation for intestinal adhesion was performed 4 years later. At presentation, he sat with a pulsating internal jugular vein rising up to the mandible angle. Also, a palpable right ventricular heave, a loud P2 and a GrIII/VI pansystolic murmur at the left sternal border were heard. The abdomen was distended with shifting dullness indicative of ascites, and both legs were edematous. Furthermore, a continuous bruit was heard over the left paraumbilical area. The electrocardiogram showed atrial fibrillation with right ventricular hypertrophy. The transthoracic echocardiography revealed moderate tricuspid regurgitation (pressure gradient 52 mmHg) and pulmonary hypertension, and the left ventricular ejection fraction was 83%. The pulmonary function test and lung ventilation-perfusion scan results were both unremarkable. Initially the contrast-enhanced abdominal computed tomographic angiography and magnetic resonance angiography (MRA) revealed a high-flow arteriovenous shunting originated from the left renal area with marked engorgement of the left renal vein (36 mm wide) and inferior vena cava (IVC) (40 mm wide) (Figure 1). Subsequently, dynamic abdominal spiral computed tomography (spiral CT) with 3D volume rendering confirmed numerous arteriovenous (AV) fistulae around the atrophic left kidney. Feeding arteries with mild to moderate dilation came from the splenic artery, the left renal artery, translumbar arteries and the renal capsular branch of the left inferior phrenic artery (Figure 2). Cardiac catheterization demonstrated a step-up of oxygen saturation from the low (77%) to high IVC (92%) level, a high cardiac output (8.62 L/min, index 5.7 L/min/m2) and moderate pulmonary hypertension (pulmonary artery pressure, 69/26 mm Hg, mean 40 mm Hg). Due to complexity of the high-flow AV fistulae associated with
long-standing pulmonary hypertension, conservative treatment including oral sildenafil was then adopted. The patient’s clinical condition remained stable for around 16 months till the preparation of this manuscript.

DISCUSSION

Secondary pulmonary hypertension due to systemic AV fistula has been recognized in various settings. These include created AV fistula for chronic dialysis, spinal disc surgery, various forms of trauma, etc. In a review of 12 cases of traumatic renal AV fistula, a majority (11 cases) presented with gross hematuria. None of them presented with heart failure or pulmonary hypertension. To our knowledge, it is rare for peri-renal AV fistulae to present with pulmonary hypertension and right-side heart failure.

Etiologically, the renal AV fistulae can be classified into (1) congenital, (2) idiopathic, (3) in association with renal cell carcinoma, (4) post-nephrectomy, (5) following renal trauma. This patient had a past history of abdominal penetrating injury about 16 years ago. From the image study, the renal AV fistula was not cirrhotic or angiomatous in appearance, which is the typical presentation of congenital renal AV fistula. So, the etiology of this peri-renal AV fistula was most likely from penetrating trauma.

The pathologic basis and the pathogenesis of the AV fistula leading to pulmonary hypertension are yet to be clarified. The pulmonary vascular bed is highly distensible initially and compensates for the increased pulmonary blood flow. However, the enhanced pulmonary blood flow produces mechanical vascular strain, compromising pulmonary vascular bed compliance. Progressive pulmonary vascular remodeling with elevated pulmonary arterial pressure could result.

Experimentally, increase in collagen content and wall thickness of pulmonary arteries, and extension of muscle into smaller pulmonary arteries, were demonstrated in created AV fistula in the fetal lamb. In patients who receive chronic hemodialysis via AV fistula presenting with pulmonary hypertension, the high blood flow and the altered vascular tone due to imbalance between the vasodilator (nitric oxide, prostanoids) and vasoconstrictor (endothelin and thromboxane) explain it. Zaid Abassi et al. demonstrated that patients under chronic hemodialysis with pulmonary hypertension had

![Figure 1](image1.png)

**Figure 1.** Contrast-enhanced thoraco-abdominal MRA in the arterial phase (abdominal aorta, Ao, is opacified). Early opacification of the dilated left renal vein (LRV) indicates high-flow arterio-venous shunting. Inferior vena cavae (IVC) is markedly dilated, too.

![Figure 2](image2.png)

**Figure 2.** Dynamic contrast-enhanced abdominal spiral CT with 3D volume rendering in the arterial phase (abdominal aorta, Ao, is opacified). Arrows indicate the peri-renal arterio-venous fistulae. The feeding arteries are also marked. The left kidney is atrophic. Left anterior oblique projection. Abbreviations: translumbar artery, L; capsular branch of inferior phrenic artery, C; splenic artery, P; left renal artery, LRA.
lower basal nitric oxide level and lower hemodialysis-induced nitric oxide production than those under chronic hemodialysis without pulmonary hypertension. Additionally, the endothelin-1 plasma concentration in ESRD patients with or without pulmonary hypertension both showed significantly higher level than normal individuals. Whether the findings on ESRD patients with AV fistula could apply to other AV fistula with pulmonary hypertension is not known yet.

Treatment of AV fistula-related pulmonary hypertension includes surgical ligation or embolization of the culprit AV fistula if feasible. However, after discussion with the urologist and interventional radiologist, the patient declined either approach due to the potential procedural risks. Currently, we have instituted sildenafil, a phosphodiesterase-V inhibitor, in addition to diuretics for his pulmonary arterial hypertension, and his clinical symptoms and severity of pulmonary hypertension have remained stable for 5 years. Takeshi Nara et al. have administered prostaglandin E and phosphodiesterase III inhibitor in AV fistula-related biventricular failure after closure of the AV fistula perioperatively. In such cases, abrupt closure of the fistula could increase afterload, decrease cardiac output, and increase wedge pressure. Prostaglandine E or phosphodiesterase III inhibitor improved the cardiac index and reduced pulmonary artery pressure.

In conclusion, we have presented a case with the initial presentation of pulmonary hypertension and right-side heart failure secondary to AV fistula caused by abdominal stab injury 16 years ago. AV fistula is an easily ignored but potentially reversible cause of pulmonary hypertension. In some cases, medical treatment remains the last effective option if interventional approaches are not adopted.

REFERENCES