Of a Tortuous Nature

An Unusual Cause of Peripartum Congestive Heart Failure

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Information about a real patient is presented in stages (boldface type) to expert clinicians (Drs Rubin, Grinspan, and Ginns), who respond to the information, sharing their reasoning with the reader (regular type). A discussion by the authors follows.

Patient presentation: A 42-year-old recently postpartum, multiparous woman presented to the emergency department with dyspnea on exertion. Full-term pregnancy and delivery 11 days earlier had been uncomplicated. Five days before admission the patient experienced progressive exercise intolerance, fatigue, and peripheral edema associated with chest discomfort and palpitations. On the evening before presentation, she developed paroxysmal nocturnal dyspnea. Her other medical and surgical history was unremarkable. Obstetric history was notable for 11 pregnancies, with 10 uncomplicated spontaneous vaginal deliveries and 1 prior miscarriage. She was not prescribed any home medications, denied allergies, and had no toxic habits. There was no family history of cardiac or clotting disorders. She denied syncope, fever, cough, sore throat, rash, bleeding, and flank pain.

Dr Ginns: The differential diagnosis of postpartum dyspnea is broad and includes syndromes specifically associated with pregnancy and common syndromes unmasked by the peripartum state of both cardiovascular and noncardiovascular etiologies (Table).[1] The story of progressive dyspnea, edema, and orthopnea suggests a heart failure syndrome. The patient's increased age and multiparity place her at higher risk for peripartum cardiomyopathy, which may manifest toward the end of pregnancy or within months thereafter. It will be helpful to investigate the peripartum transfusion and tocolytic requirements, which are associated with the development of pulmonary edema. Acute myocardial ischemia and pulmonary embolism must be ruled out.

***Patient presentation (continued)*: In the emergency department, the patient appeared well at rest. Her heart rate was 96 and blood pressure was 148/79 mm Hg. Oxygen saturation was 97% at rest, but, on ambulation, it decreased to 89% with associated tachypnea. The body mass index was 29 kg/m^2 and the body surface area was 1.52 m^2. Jugular veins were distended with an estimated central venous pressure of 14 cm H_2O. Cardiac examination revealed tachycardia without murmurs or gallops. Lung sounds were diminished at the bases. It is noteworthy that auscultation of the left midlung field revealed a high-pitch continuous murmur. The murmur was also present in the epigastrium, but not well appreciated over the precordium. The abdominal examination furthermore revealed hepatomegaly and a palpable spleen tip. The abdomen was soft and nontender, and there were no signs of ascites. Extremities were warm, with moderate pitting edema and intact distal pulses. There was no joint swelling or skin findings.**

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Table. Differential Diagnosis of Postpartum Dyspnea

Diagnosis	Symptoms
Cardiovascular syndromes associated with pregnancy	Heart failure attributable to peripartum cardiomyopathy or dilated cardiomyopathy
	Myocardial infarction attributable to spontaneous coronary artery dissection, atherosclerotic plaque rupture, or coronary embolism
	Venous thromboembolism, pulmonary embolism, amniotic fluid embolism
	Mitral valve regurgitation
	Arrhythmias: premature depolarization, ventricular premature depolarization, supraventricular tachycardia
	Tocolytic-induced pulmonary edema
	Aortic dissection and systemic arterial dissection
Noncardiovascular syndromes associated with pregnancy	Postpartum hemorrhage
	Preeclampsia
	Reactive airways disease
	Infection/pneumonia
	Thyroid disease
	Rheumatologic vasculitis
	Iatrogenic volume overload

Abnormal basic laboratory tests included N-terminal probrain natriuretic peptide 1501 pg/mL (reference, 0–150 pg/mL), D-dimer 2.16 μg/mL (reference, <0.5 μg/mL), and alkaline phosphatase 190 IU/L (reference, 44–147 IU/L). Otherwise, blood work was normal, including hemoglobin, platelets, creatinine, albumin, prothrombin time, thyroid-stimulating hormone, creatinine kinase, and troponin. Admission ECG showed sinus tachycardia without injury current. A chest x-ray revealed pulmonary edema without frank consolidation.

Dr Ginns: The patient's physical examination and blood work are consistent with decompensated left- and right-sided heart failure. The absence of ECG changes and troponin elevation lowers the suspicion for an acute coronary syndrome attributable to thrombosis, vasculitis, or dissection. Without other findings, the suspicion for peripartum cardiomyopathy would be high. Yet in this case, the unexpected continuous abdominal murmur and hepatosplenomegaly raise the prospect of a rare etiology for the patient's decompensation. Pathological continuous murmurs indicate blood flow from a high-pressure system to a low-pressure system such as occurs in patent ductus arteriosus (PDA) or coronary-cameral fistula, or are alternatively attributable to a severe fixed arterial stenosis such as aortic coarctation. A PDA or coronary fistula associated with significant volume load would likely have manifested itself earlier than the 11th pregnancy. A coarctation murmur would not transmit to the upper abdomen and would have been associated with diminished lower limb pulses.[1] The patient's extrathoracic murmur raises the possibility of an arteriovenous fistula (AVF).

***Patient presentation (continued)*: Computed tomography (CT) angiogram of the chest and upper abdomen revealed a moderately enlarged pulmonary artery without emboli and bilateral pleural effusions. The abdomen showed hepatomegaly and marked dilatation of the splenic vein, portal vein, hepatic veins, splenic artery, and celiac artery (Figure 1). Transthoracic echocardiogram showed a normal left ventricular ejection fraction with mildly enlarged ventricles. The estimated pulmonary artery systolic pressure was 59 mm Hg. There was no valvular disease, atrial septal defect, PDA, or coarctation.**

Dr Ginns: In summary, this previously healthy postpartum patient presenting with dyspnea is discovered to have pulmonary edema with preserved left ventricular ejection fraction and right ventricular (RV) dilation with pulmonary hypertension. In the setting of a continuous murmur on examination and the tortuous ectatic vasculature by imaging, there is concern for an AVF causing augmentation of venous return and resultant heart failure. A catheterization of the right side of the heart with oximetric recordings is indicated.

***Patient presentation (continued)*: The patient was transferred to the cardiac care unit and received treatment with intravenous furosemide. The on-call fellow performed a catheterization of the right side of the heart overnight. The heart rate was 92 and the blood pressure was 143/74. The procedure revealed the following hemodynamics: right atrium (RA) 14 mm Hg, RV 43/6 mm Hg, pulmonary artery 43/20 mm Hg (mean 28 mm Hg), and mean pulmonary capillary wedge 18 mm Hg. A limited saturation run was performed that revealed the following: superior vena cava 54%, RA 76%, RV 82%, pulmonary artery 84%. The arterial oxygen saturation was 97%. Cardiac output was calculated as 9.0 L/min by the Fick method and systemic vascular resistance was 780 dynes·s^{-1}·cm^{-5}. The cardiac index was 5.92 L·min^{-1}·m^{-2} (normal value, 2.5–4.0 L·min^{-1}·m^{-2}) and the Qp:Qs was 3.3:1.**

Dr Ginns: The catheterization confirms the presence of decompensation in the right side of the heart with mild pulmonary hypertension and a high-output state with a left-to-right shunt.

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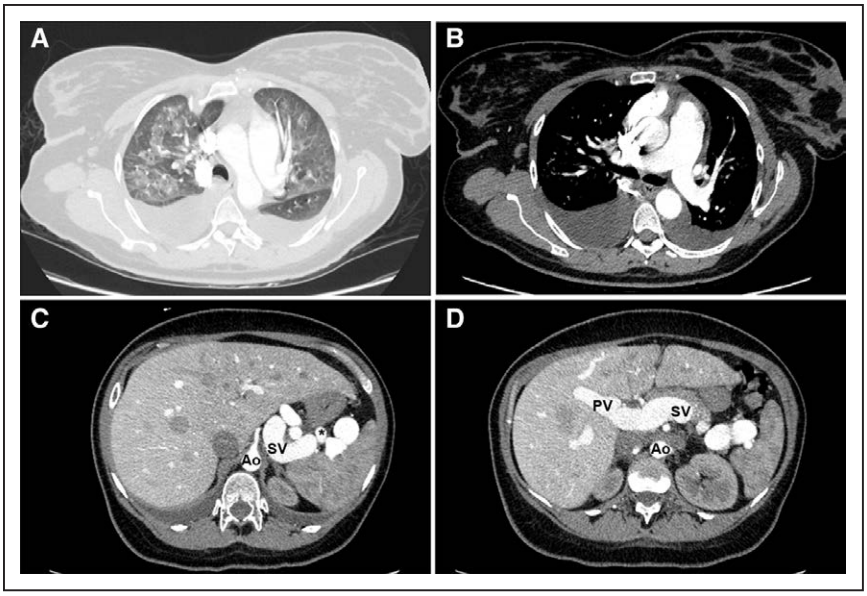

Figure 1. Computed tomography angiogram of the chest and upper abdomen showing ground-glass opacities consistent with pulmonary edema and moderate-sized pleural effusions (A), and a dilated main pulmonary artery without evidence of pulmonary embolism (B). Abdominal computed tomography (C and D) reveals hepatosplenomegaly, and dilated splenic artery (asterisk), splenic vein (SV), and portal vein (PV), as well. Note the relatively small-sized descending aorta (Ao) in comparison with the enlarged splenic vein.

The Qp:Qs ratio represents the size of a shunt by quantifying the amount of blood flow through the pulmonary circulation relative to the systemic circulation. It is derived from an oximetry run by dividing the oxygen content of pulmonary blood by the oxygen content of systemic blood using the following modified formula: Qp:Qs = ($O2sat_{Arterial}$-$O2sat_{SVC}$) / ($O2sat_{PV}$-$O2sat_{PA}$), where PV indicates pulmonary vein; PA, pulmonary artery; and SVC, superior vena cava. The pulmonary vein saturation is considered identical to the arterial oxygen saturation if there is no concern for right-to-left shunt. Clinically, a left-to-right shunt causing Qp:Qs>2:1 requires invasive treatment.

The size of the oxygenation step-up between the superior vena cava and the pulmonary artery (30%) was larger than the normal range between 2 right-sided cardiac chambers (<8%), indicating the presence of a shunt at the atrial, ventricular, or great vessel level.[1] More specifically, in the setting of an oxygenation step-up from the vena cava to the RA, the differential diagnosis includes atrial septal defect, ventricular septal defect with concomitant tricuspid regurgitation, coronary-cameral fistula, partial anomalous pulmonary venous return, PDA, and AVF occurring below the diaphragm. Resources were limited at the midnight hour of the patient's admission, but ideally a complete oximetry run would have been performed with blood sampling from the inferior vena cava and various levels of the cardiac chambers to localize the shunt. Nevertheless, there are sufficient data to synthesize a working diagnosis.

An atrial septal defect causes an oxygenation step-up between the vena cava and the right atrium, but is unlikely to cause a loud murmur. A ventricular septal defect was not localized on the echocardiogram, and the initial oxygen step-up would occur in the RV, not the RA, and the murmur would not be continuous. A coronary-cameral fistula to the RA or RV might be associated with a large shunt and would be visualized on echocardiogram as an atypically large coronary artery, or by the high-velocity jet entering a right-sided cardiac chamber on color Doppler. Scimitar syndrome, an anomalous pulmonary vein running from the right lung to the inferior vena cava, would produce an initial oxygenation step-up in the inferior vena cava, but would be associated with isolated right-sided heart failure instead of left-sided heart failure with pul-

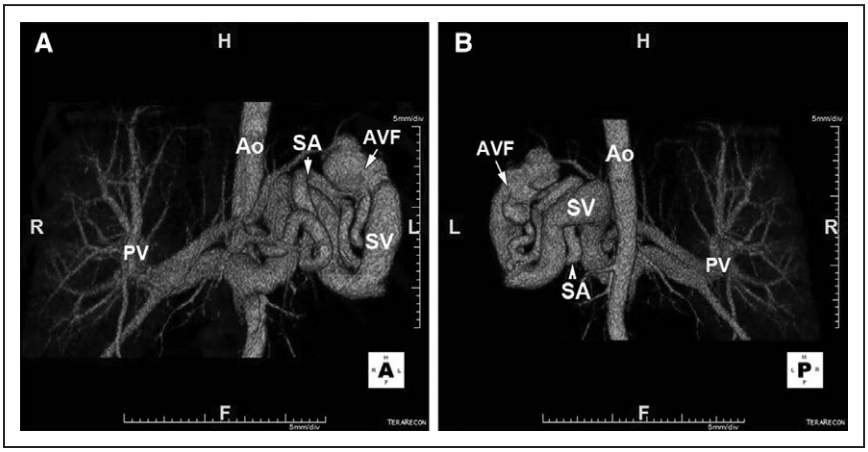

Figure 2. Three-dimensional reconstruction of arteriovenous fistula (AVF) and tortuous splanchnic vasculature from computed tomography angiogram is shown in the anterior view (A) and posterior view (B). Ao indicates aorta; SA, splenic artery; SV, splenic vein; and PV, portal vein.

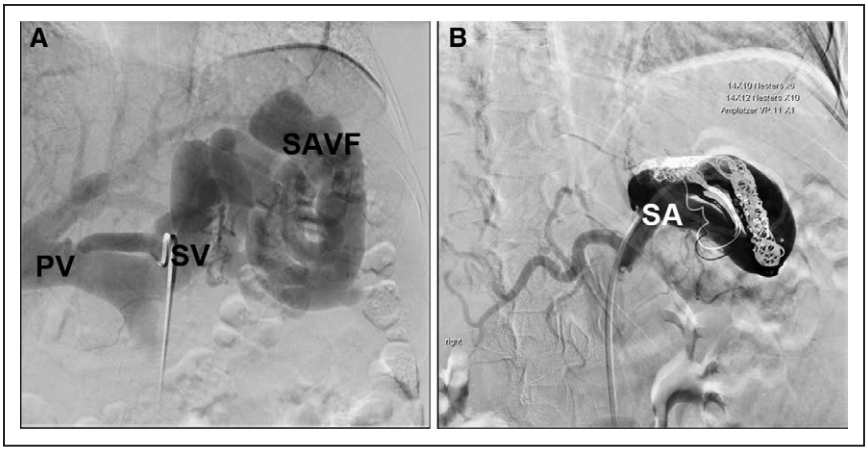

Figure 3. Angiography and coiling of splenic artery.
A, Initial contrast angiography of celiac artery plexus demonstrates splenic arteriovenous fistula (SAVF) with a dilated splenic vein (SV) and portal vein (PV). **B,** Angiography of the splenic artery (SA) after placement of coil and successful embolization demonstrates no further contrast filling the venous system.

monary edema. A PDA would have produced an oxygen saturation step-up in the pulmonary artery and would not cause an elevated RA or RV saturation unless there was significant pulmonary and tricuspid insufficiency.

The abnormally enlarged splenic vessels seen on CT suggest localization of the shunt to an infradiaphragmatic AVF. The presence of this type of fistula will lead to an increased volume load on both ventricles and high-output heart failure. In this patient, high-output heart failure is felt to be attributable to probable rupture of a small, previously unrecognized chronic splenic artery aneurysm (SAA) causing a high-flow splenic artery-splenic vein fistula. To treat the patient's heart failure, the shunt will have to be closed either percutaneously or surgically.

***Patient presentation (continued)*: Consultation with the liver transplant team was obtained for further evaluation of hepatomegaly without other signs of portal hypertension. If liver fibrosis were present, the risk for portal vein thrombosis with a percutaneous fistula closure would be high because of the stasis of flow into the fibrotic liver after occlusion of the vascular bed.[2] A transjugular liver biopsy revealed mild sinusoidal congestion without fibrosis. The hepatic venous pressure gradient was 3 mm Hg, indicating the absence of portal hypertension. The patient underwent CT angiography, which confirmed the presence of a fistula between the splenic artery and splenic vein (Figure 2). The patient underwent invasive angiography and the arteriovenous communication was successfully embolized with Nester coils and an Amplatzer vascular plug II device (Figure 3). The procedure was followed by flank pain attributable to large splenic infarcts and systemic hypertension, which was felt to be caused by a sudden rise in systemic vascular resistance. Transthoracic echocardiogram revealed new mild reduction in left ventricular ejection fraction and a decrease in LV size. β-Blocker and angiotensin-converting enzyme inhibitor were initiated. Repeat CT abdomen before discharge showed infarction of the spleen and absence of portal vein thrombosis (Figure 4). She was prophylactically given meningococcal and pneumococcal vaccinations and then discharged home 7 days after the procedure.**

DISCUSSION

The patient's final diagnosis was high-output heart failure attributable to a large AVF between the splenic artery and splenic vein, the first such case described in the literature.

The association between AVF and high-output heart failure was first reported in the early 1970s.[3] Formal diagnosis of high-output heart failure may be confirmed with a catheterization of the right side of the heart re-

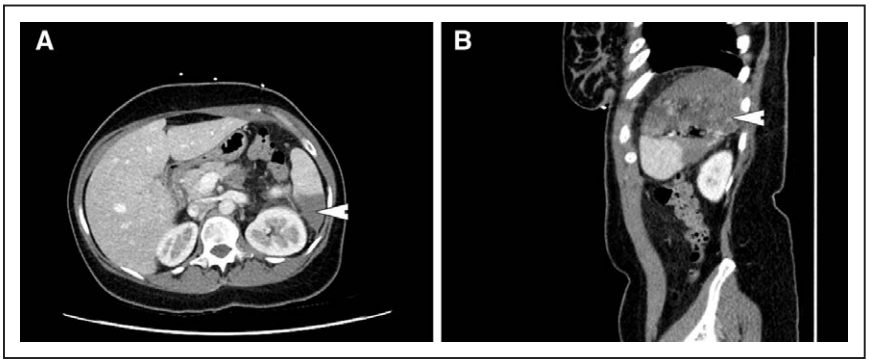

Figure 4. Computed tomography abdomen pelvis postembolization in axial (A) and sagittal (B) views showing hypodense area of infarcted spleen (white arrow).

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vealing elevated filling pressures with high cardiac output indexed for body size (cardiac index >4 $L \cdot min^{-1} \cdot m^{-2}$ with cardiac output >8 L/min) and low systemic vascular resistance (< 800 $dynes \cdot s^{-1} \cdot cm^{-5}$). The diagnosis of AVF in this case is facilitated by an oximetry run performed in the catheterization laboratory. A sample from the inferior vena cava or hepatic vein, the closest accessible vascular structures to the abdominal AVF, would have revealed a nearly arterial oxygen saturation and further localized the shunt to the portal vasculature.

Although the Qp:Qs derived from an oximetry run reflects the magnitude of shunting, in the instance of a peripheral extracardiac shunt, the patient's unique anatomy must be considered. Simply deriving the Qp:Qs calculation as 3.3:1 would lead the diagnostician to believe that there is a >3-fold greater volume load on the right side of the heart than on the left side of the heart. In this case, however, the volume load affects both sides of the heart. Additional flow from an extracardiac AVF is transmitted through both the pulmonary and systemic circulations, producing both right and left ventricular dilatation, with eventual left-sided heart failure causing pulmonary edema and right-sided heart failure causing hepatomegaly.

An AVF specifically involving the splenic vasculature is a rare congenital or acquired shunt in the setting of an SAA. SAA is the most common visceral arterial aneurysm, and occurs 4 times more frequently during pregnancy. The condition most commonly affects multiparous women (mean 4.5 pregnancies patient in 1 small study).[4]

The unique combination of molecular and physiological changes during pregnancy contribute to the development of SAAs by ultimately weakening the arterial wall and causing aneurysmal dilatation.[5] On a molecular level, the hormones estrogen, progesterone, and relaxin have been linked to internal elastic lamina fragmentation, medial degeneration, and enhanced arterial elasticity. Increased renin and aldosterone also contribute to arterial wall thinning in the volume-expanded state. From a hemodynamic perspective, the physiological increase of cardiac output, blood pressure, portal blood flow, and shear stress predispose the splenic vasculature to aneurysm formation and rupture. Frank SAA rupture during pregnancy is usually fatal and carries a 75% mortality rate.

CONCLUSION

The case challenged the traditional differential diagnosis for new-onset congestive heart failure and highlighted the importance of integrating the physical examination, imaging, and catheterization, which all played a role in uncovering the diagnosis and directing management.

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DISCLOSURES

None.

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FOOTNOTES

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