Case Reports

Venous Myocardial Infarction in an Infant

with Obstructed Totally Anomalous Pulmonary Venous Drainage and Coronary Sinus Ostial Atresia

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We report a rare causal association between obstructed supracardiac totally anomalous pulmonary venous drainage and coronary sinus ostial atresia. Our 12-week-old patient developed venous myocardial infarction secondary to coronary venous hypertension because her sole route of coronary venous drainage was obstructed. She recovered after the obstruction was relieved by balloon dilation. Surgical repair then included anastomosis of the pulmonary venous confluence to the left atrium, ligation of the vertical vein, and unroofing of the coronary sinus. Coronary sinus ostial atresia is rarely diagnosed before autopsy. ***(Tex Heart Inst J 2016;43(5):430-2)***

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Coronary sinus (CS) ostial atresia is a rare entity, known to be associated with a variety of congenital heart diseases.[1-6] Generally, an anomalous decompressing vein drains the coronary venous blood. Any obstruction in the path of coronary venous drainage can lead to coronary venous hypertension and myocardial infarction (MI). We report a case of obstructed totally anomalous pulmonary venous (TAPV) return with CS ostial atresia, which led to venous MI. This, to our knowledge, has never before been reported.

Case Report

In March 2013, a 12-week-old girl with a history of failure to thrive presented in extremis with cardiogenic shock. Her physical examination showed that she had tachypnea, poor systemic perfusion, cyanosis, and bibasilar rales. An emergency echocardiogram revealed a membranous obstruction at the junction of the brachiocephalic vein and the superior vena cava (Fig. 1) and TAPV drainage of a supracardiac type—together with a dilated CS, a large atrial septal defect with right-to-left shunting, and pulmonary hypertension.

Emergency cardiac catheterization, performed to relieve the obstruction, confirmed the presence of TAPV drainage through a vertical vein into the left brachiocephalic vein, which was obstructed at its junction with the right superior vena cava (gradient, 19 mmHg). This CS ostial atresia resulted in severe pulmonary hypertension and retrograde drainage, via a connecting vein, to the pulmonary venous confluence (Fig. 2). Balloon angioplasty at the site of the obstructive membrane substantially relieved the hypertension.

During the procedure, the patient developed sustained monomorphic ventricular tachycardia secondary to an MI, as evidenced by troponin levels elevated to 130 ng/mL and by diffuse ST-segment changes on electrocardiogram. The infarction was most likely associated with the transient elevation of coronary pressure upon balloon inflation.

The patient was noted on her echocardiogram to have global qualitative left ventricular dysfunction, and her myocardial function recovery was closely observed in the intensive care unit. Her cardiac enzyme levels returned to baseline, and her left ventricular function improved substantially. A cardiac computed tomogram, obtained a week later to better delineate the postoperative pulmonary venous anatomy, confirmed the above findings (Fig. 3).

Two weeks later, the patient underwent surgical repair, which comprised anastomosis of the venous confluence to the left atrium, unroofing of the CS to the left atrium, ligation of the vertical vein and the connecting vein from the CS to the confluence,

and closure of the atrial septal defect with a fenestrated patch. On follow-up evaluation, we noted a gradient of 6 mmHg at the confluence, with left-to-right shunting across the fenestration. She was discharged from the hospital 3 weeks after surgery. Postoperatively, she continued to experience pulmonary hypertension, which necessitated sildenafil and oxygen therapy at home.

Discussion

We report a rare case of obstructed supracardiac TAPV drainage in the presence of CS ostial atresia, leading to MI from coronary venous hypertension. Coronary sinus ostial atresia is a rare entity known to be associated with a variety of congenital heart diseases.[1-6] Until now, its association with obstructed TAPV drainage has not been reported.

In the presence of CS ostial atresia, the CS blood might drain into the left superior vena cava and eventually return to the right atrium (having taken a longer route) or return directly into the cardiac chambers.[5] Any obstruction in the sole path of CS drainage will increase pressure in the coronary venous circuit, leading to intramyocardial edema and necrosis. Several cases have been reported[2,4,6] of myocardial congestion and ischemia as a result of coronary venous hypertension from inadvertent ligation of left superior venae cavae in patients with CS

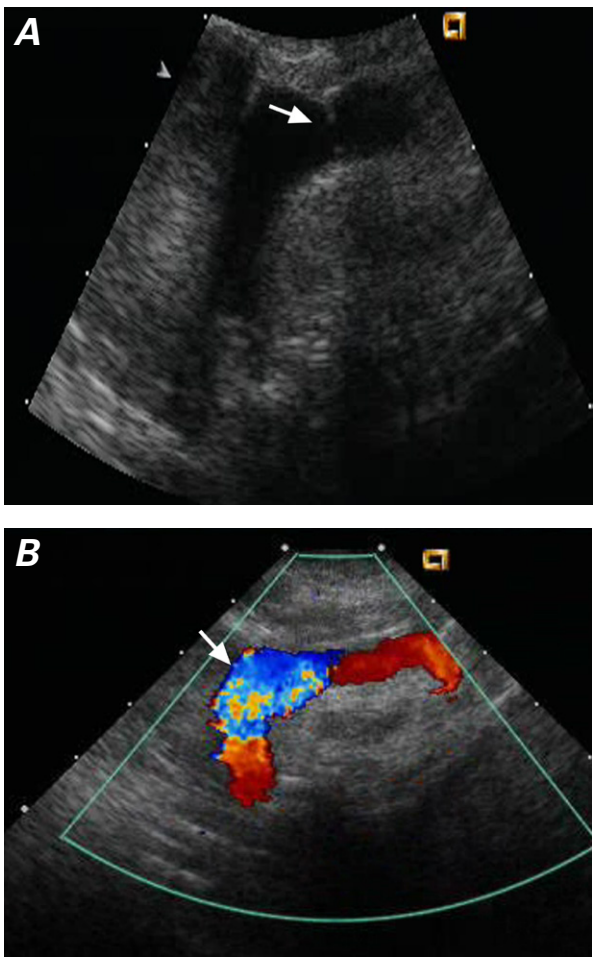

Fig. 1 *Transthoracic echocardiograms show **A**) an obstructive membrane (arrow) at the junction of the brachiocephalic vein and right superior vena cava and **B**) flow acceleration (in color-flow Doppler mode) at the site of membranous obstruction (arrow).*

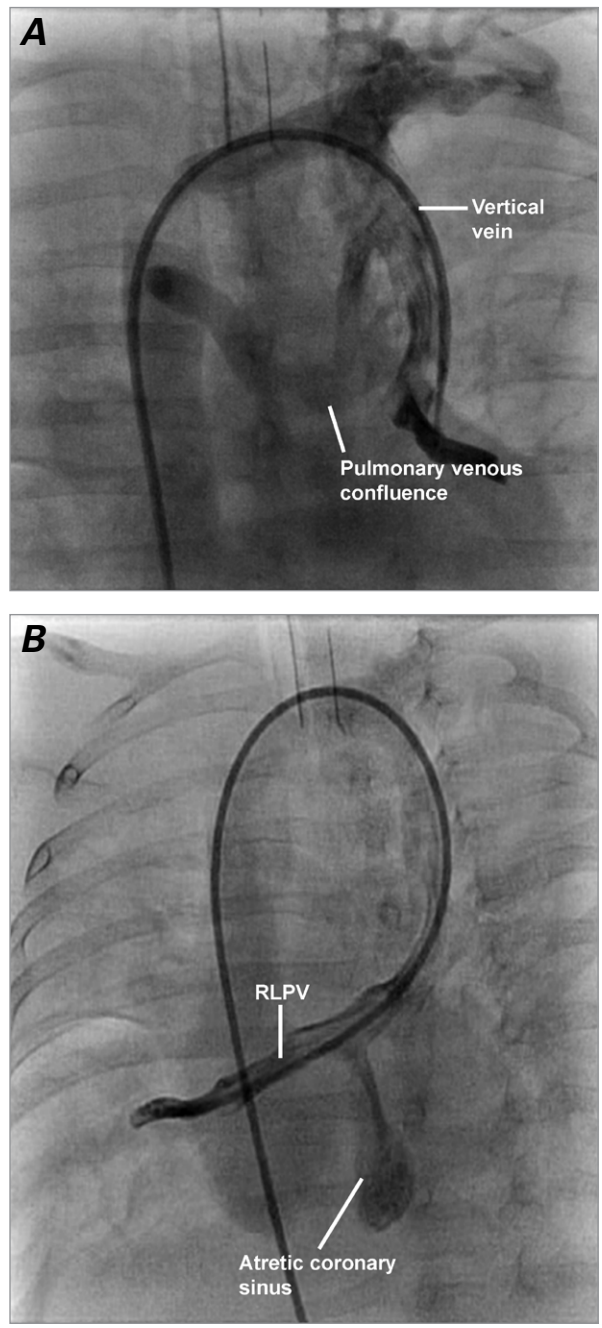

Fig. 2 *Angiograms show **A**) pulmonary venous confluence draining into the vertical, brachiocephalic, and right superior caval veins; and, **B**) a catheter in the right lower pulmonary vein (RLPV), atresia of the coronary sinus ostium, and a connecting vein from the coronary sinus to the pulmonary venous confluence.*

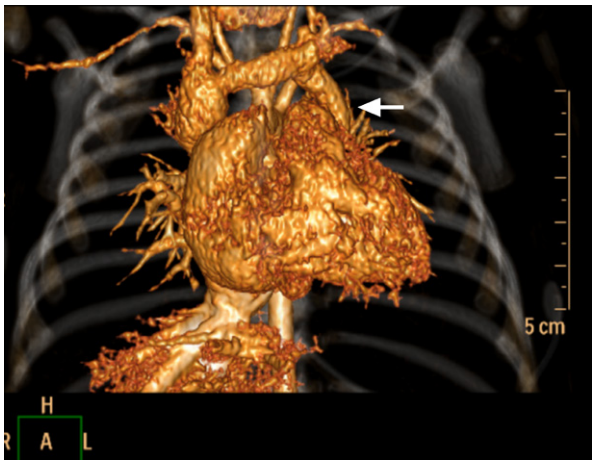

Fig. 3 *Cardiac computed tomogram (3-dimensional reconstruction) shows totally anomalous pulmonary venous drainage from the vertical vein (arrow) into the brachiocephalic vein.*

ostial atresia. Our patient was unstable at presentation because of obstructed TAPV drainage, and possibly because of myocardial congestion and ischemia from coronary venous hypertension. Once the obstruction was relieved, her clinical status substantially improved.

Coronary sinus ostial atresia is suspected when a prominent CS and a reversal of flow are seen on echocardiograms via color-flow Doppler echocardiography. However, diagnosis is generally confirmed by angiography, which can clearly reveal an atretic CS ostium and retrograde flow into the brachiocephalic vein.[1,2,4,7,8] Multidetector computed tomography and cardiac magnetic resonance have also proved useful in evaluating the CS anatomy and drainage.[5] The importance of fast and accurate delineation of the CS anatomy cannot be overstated, because a delay in diagnosis means incurring a risk of MI.

The reported surgical techniques for treating CS ostial atresia include unroofing the CS to the left atrium[8] or creating a large opening between the CS and the left atrium, via a left superior vena caval flap.[9] In a patient with unobstructed TAPV drainage and CS ostial atresia, there was a report[1] of dissection of the vertical vein and anastomosis of that vein to the left atrium, together with anastomosis of the connecting vein from the CS to the left atrial appendage. In our patient, the connecting vein was transected and the CS was unroofed in its entirety in the left atrium, along with transection of the vertical vein and anastomosis of that vessel to the left atrium.

Coronary sinus ostial atresia is rarely diagnosed before autopsy. In a living patient, we report a rare causal association between obstructed supracardiac totally anomalous pulmonary venous drainage and CS ostial atresia.

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