Partial Anomalous Hepatic Venous Return: A Systematic Intraoperative Exclusion of Other Serious Diagnoses

Camellia D. Asgarian, MD,* Susan M. Martinelli, MD,* Jonathan B. Mark, MD,† and Priya A. Kumar, MD‡

A 83-year-old gentleman with a remote history of myocardial infarction presented with a witnessed cardiac arrest secondary to ventricular tachycardia. The patient was resuscitated successfully and, after coronary catheterization, was found to have severe coronary artery disease. A transthoracic echocardiogram (TTE) reported an ejection fraction of 40% to 45%, left ventricular hypertrophy, biventricular enlargement, and a possible patent foramen ovale. In addition, the patient had a ventricular endocardial pacemaker in place for atrial fibrillation and bradycardia. The patient was scheduled for 3-vessel coronary artery bypass grafting. After an uneventful induction of anesthesia, transesophageal echocardiography (TEE) showed an ejection fraction of 25% to 30%, with severe hypokinesis in the inferior, inferoseptal, anteroseptal, and anterior walls, biventricular enlargement, and mild mitral and tricuspid regurgitation. A patent foramen ovale was excluded via color flow Doppler (CFD) and an IV agitated saline study. Verbal informed consent was obtained from the patient via telephone for publication of this case report and any accompanying images.

Closer examination of the midesophageal right ventricular inflow-outflow view showed an aberrant opening in the free wall of the right atrium (RA). CFD interrogation of this opening demonstrated a vascular flow entering the RA from the free wall and directed toward the atrial septum (Fig. 1A; Supplemental Digital Content 1, Supplemental Video 1, http://links.lww.com/AA/B272). Pulsed wave Doppler interrogation demonstrated a low-velocity laminar venous flow pattern with an antegrade diastolic flow peak (Fig. 1B). An IV agitated saline contrast study performed via the right internal jugular vein central line showed the bubbles appearing from the superior vena cava (SVC), which were then cleared by the flow in question, consistent with negative echo contrast (Fig. 2; Supplemental Digital Content 2, Supplemental Video 2, http://links.lww.com/AA/B273). Normal pulmonary venous flow was observed entering the left atrium. A second IV contrast study with agitated saline injected via a lower extremity vein was performed. The bubbles emerged from the expected location of the inferior vena cava (IVC) and not from the SVC or the aberrant vessel in question, confirming that the vessel was not an aberrant IVC. This also excluded the diagnosis of total anomalous hepatic venous return. Having excluded the earlier possibilities in our patient, the surgeon performed a thorough inspection noting no obvious additional vascular abnormalities. Standard venous cannulation with a dual-stage cannula was then performed via the right atrial appendage, and the cannula tip was advanced into the IVC without difficulty. A diagnosis of exclusion was made that the vessel in question was an aberrant hepatic vein draining directly into the RA. The remainder of the surgical course and recovery was uneventful. A focused TTE was performed postoperatively, which traced this aberrant vessel into the liver, thus confirming the earlier diagnosis. The patient’s postoperative course was uneventful, and he was doing well 1 year after surgery.

DISCUSSION

When faced with an aberrant vessel inserting into the free wall of the RA, several concerning differential diagnoses need to be excluded.

Coronary artery fistulas are rare defects that affect approximately 0.2% of the adult population. They are usually congenital but can be caused by infection, trauma, or surgery. They generally arise from the right coronary artery, terminate in the RA or right ventricle, and are associated with an abnormally dilated coronary artery. This was excluded in our patient because there was no abnormal dilation in the coronary arteries visualized on TEE. CFD can be used to identify coronary artery fistulas, which would show continuous turbulent flow during systole and diastole. In our case, a laminar predominantly diastolic venous flow pattern excluded this anomaly. In addition, neither cardiac catheterization nor surgical inspection reported any such anomalous coronary artery connection.

In 2011, Kuroda et al. described the anomalous insertion of the IVC into the RA. The IVC typically inserts into the RA inferiorly in a longitudinal axis with the SVC. With this anomaly, the IVC connects to the RA laterally, causing an abnormal angulation. Venous cannulation for cardiopulmonary bypass can be complicated by an aberrant IVC, in that the cannula can perforate the vein or terminate in the right hepatic vein. On TEE examination, after obtaining the midesophageal bicaval view, one can trace the IVC inferiorly to the level of the liver where the hepatic veins can be visualized draining into the IVC. A second IV contrast study with agitated saline injected via a lower extremity vein can be performed to identify the IVC drainage. In addition, surgical inspection should reveal the insertion site of the IVC.
Partial anomalous pulmonary venous return is estimated to have a prevalence of 0.4% to 0.7% in the adult population by autopsy report. Patients can present with shortness of breath, heart murmur, supraventricular arrhythmia, or decreased exercise tolerance. Approximately 12.1% of these anomalies have aberrant connections to the RA or RA-SVC junction as shown in Figure 3A. An absence of the pulmonary vein insertion into the left atrium would support this diagnosis. Furthermore, the vast majority of partial anomalous pulmonary venous return in adults is seen in association with a sinus venosus type atrial septal defect, which was excluded in the subject patient of this case report.

Total anomalous hepatic venous return is also known as infrahepatic interruption of the IVC. In this case, the infrahepatic portion of the IVC empties into the azygos system, which inserts into the SVC. The hepatic veins form a single confluence and empty directly into the RA (Fig. 3B). This diagnosis can mimic aortic pathology because the azygos vein runs parallel to the aorta and is usually dilated in these patients, thus potentially causing the misdiagnosis of aortic dissection. Also, surgical ligation of the azygos vein, as is occasionally done in some abdominal surgeries, can be fatal if this aberrancy is not recognized. This diagnosis can be confirmed by injecting agitated saline into a lower extremity vein and visualizing the bubbles entering the RA via the SVC. This was excluded in our patient.

Partial anomalous hepatic venous drainage is relatively common in the pediatric population in patients with other cardiac anomalies. D'Cruz and Smith described the first 2 adult cases of direct drainage of a hepatic vein into the RA. The diagnosis was made with the subcostal view on TTE. The aberrant vessel was associated with an “extra” eustachian valve in both cases and was of no clinical significance other than creating an abnormal echocardiogram. Figure 3C illustrates normal venous anatomy where the hepatic veins join the IVC below the diaphragm. The circled area in the figure demonstrates the abnormality shown in this report, where one hepatic vein may diverge and enter the RA directly. Having excluded other possibilities in our patient, a focused TTE was performed postoperatively, which traced this aberrant vessel into the liver, thus confirming that it was an aberrant insertion of a hepatic vein into the RA.

It should be noted that some of the conditions described earlier may have implications for line placement, particularly for pulmonary artery catheters (PAC). The abnormality we identified was an anomalous venous connection to the RA. A PAC entering the RA from the SVC or IVC could theoretically enter this anomalous vein rather than cross the tricuspid valve into the right ventricle and pulmonary artery. Antegrade flow...
from this venous connection should not favor PAC entry into the anomalous vein. However, if advancing the PAC does not record anything other than a venous pressure waveform despite insertion to 35 cm, then coiling in the RA or entry into the anomalous vessel should be suspected.

The clinical significance of partial anomalous hepatic venous return is minimal; however, alternative diagnoses (Table 1) for an aberrant vessel in the right atrial free wall can cause significant patient morbidity and potentially lead to devastating outcomes. It is important to be familiar with the differential diagnosis for this vascular anomaly, rare as some of these conditions may be, and to be able to quickly and methodically arrive at the correct diagnosis.

Figure 3. Abnormal right atrial venous connections (A) partial anomalous pulmonary venous return: The arrow shows the aberrant pulmonary veins inserting into the superior vena cava-right atrial junction. B, Infrahepatic interruption of the inferior vena cava (IVC) or total anomalous hepatic venous return. The IVC drains directly into the azygos vein, which then empties into the superior vena cava (SVC). The hepatic veins form a confluence and enter the right atrium (RA). C, Normal venous anatomy of hepatic veins joining IVC below the diaphragm. Circled area shows where one hepatic vein may diverge and enter the RA directly, as was seen in this case.
Table 1. Differential Diagnosis of Anomalous Drainage into RA

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Diagnostic features</th>
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<tr>
<td>Total anomalous PV return</td>
<td>1. Presence of PV drainage into RA, SVC, or IVC</td>
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<td></td>
<td>2. Absence of PV drainage into left atrium (LA)</td>
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<tr>
<td>Partial anomalous PV return</td>
<td>1. Aberrant PV drainage into SVC, IVC, or RA</td>
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<tr>
<td></td>
<td>2. Partial PV drainage into LA</td>
</tr>
<tr>
<td>Anomalous insertion of IVC</td>
<td>1. Aberrant vessel draining into RA</td>
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<td></td>
<td>2. Agitated contrast injection into lower extremity appears in the aberrant vessel</td>
</tr>
<tr>
<td>Total anomalous hepatic vein or infrahepatic</td>
<td>1. IVC drains into dilated azygos vein that empties into SVC</td>
</tr>
<tr>
<td>interruption of IVC</td>
<td>2. Agitated contrast injection into lower extremity appears in the SVC (not IVC)</td>
</tr>
<tr>
<td>Coronary artery fistula</td>
<td>1. Dilated coronary artery on echocardiographic examination</td>
</tr>
<tr>
<td></td>
<td>2. Cardiac catheterization demonstrates aberrant coronary drainage</td>
</tr>
<tr>
<td>Partial anomalous hepatic venous return</td>
<td>1. Diagnosis of exclusion by absence of the above features</td>
</tr>
<tr>
<td></td>
<td>2. Confirmed by surgical inspection and imaging (transthoracic echocardiogram or computerized tomography scan)</td>
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IVC = inferior vena cava; PV = pulmonary venous; RA = right atrium; SVC = superior vena cava.

DISCLOSURES

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REFERENCES

Partial anomalous hepatic venous return describes a condition in which a hepatic vein connects directly to the free wall of the right atrium (RA) rather than joining the intrahepatic inferior vena cava (IVC). The condition has minimal clinical significance although the aberrant communication may be inadvertently entered during venous cannulation for cardiopulmonary bypass.

Another variant of venous return is an anomalous connection of the IVC to the lateral RA free wall, which can typically be traced to the liver using the midesophageal bicaval view. It should be confirmed by agitated saline injection via a lower extremity vein (which would be logistically challenging during surgery). Total anomalous hepatic venous return (TAHVR), also known as interrupted IVC, represents yet another venous return variant and is characterized by diversion of the IVC into the azygos vein. Because the azygos vein empties into the superior vena cava (SVC), a lower extremity contrast injection in a patient with TAHVR will pass from the IVC into the azygos vein and then enter the RA by way of the SVC. The hepatic veins in patients with TAHVR form a confluence that drains into the RA.

In this case, pulsed wave Doppler interrogation of a color jet was noted entering the lateral RA wall consistent with venous flow. Injection of agitated saline from a lower extremity vein demonstrated the expected entry of contrast into the RA from the IVC, excluding TAHVR. Postoperative transthoracic echocardiography was able to trace the anomalous venous structure to the liver, confirming the diagnosis of partial anomalous hepatic venous return.

Correct differentiation of the various causes of anomalous flow into the lateral RA necessitates the careful use 2D Doppler and contrast echocardiography. Furthermore, echocardiographers should be familiar with the structure and function of the azygos vein, a vessel that ascends in the right hemithorax and can provide an important collateral circulation between the SVC and the IVC.