Transjugular intrahepatic portosystemic shunt with accidental diagnosis of persistence of the left superior vena cava

Ioannis Petridis, Roberto Miraglia, Gianluca Marrone, Salvatore Gruttadauria, Angelo Luca, Giovanni Battista Vizzini, Bruno Gridelli

INTRODUCTION

The transjugular intrahepatic portosystemic shunt (TIPSS) is a well known life saving procedure that is performed in patients with complications of portal hypertension, such as variceal bleeding and refractory ascites[1-5]. The standard technique for this treatment is the right internal jugular vein (RIJV) approach, although some authors have reported the left internal jugular vein (LIJV) approach when the RIJV is not available or cannot be used, due, for example, to vein thrombosis[6]. Few cases of patients with the absence of the right superior vena cava (RSVC) and persistence of the left superior vena cava was reported[7].

In the following report, we describe a rare vascular malformation characterized by the absence of the right superior vena cava (RSVC) and persistence of the left superior vena cava in a patient with a diagnosis of advanced liver cirrhosis who needed a TIPSS placement in order to control refractory ascites.

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Key words: Left superior vena cava persistence; Liver cirrhosis; Refractory ascites; Transjugular intrahepatic portosystemic shunt

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Abstract

Transjugular intrahepatic portosystemic shunt (TIPSS) is considered a valid therapeutic option for the treatment of portal hypertension and its complications. The guidelines for this procedure have already been established on the basis of the normal vascular anatomy and of various technical radiological aspects. In some rare cases, diagnosis of a congenital vascular anomaly can be made accidentally by interventional radiologists, making the procedure of the TIPSS placement extremely difficult or in some cases technically impossible. This report describes a rare vascular malformation characterized by the absence of the right superior vena cava and persistence of the left superior vena cava in a patient with a diagnosis of advanced liver cirrhosis who needed a TIPSS placement in order to control refractory ascites.

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CASE REPORT

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left superior vena cava (LSVC) have been described in the literature\(^7\). Even if this is an extremely rare situation, it can create problems during the interventional procedure. We briefly report a case in which a TIPSS was successfully created in a patient with absence of RSVC and persistence of the LSVC.

**CASE REPORT**

A 69-year-old female with a diagnosis of hepatitis C virus-related cirrhosis was referred to our institute in order to be evaluated for a TIPSS placement because of refractory ascites. In the physical examination the patient had tense ascites despite a consistent use of high dose diuretics and periodic paracentesis (every 1-2 wk). She was well oriented to time and to space and had no episodes of portosystemic encephalopathy in the past. An abdominal computed tomography (CT) scan of the abdomen and a liver Doppler sonography (US) were performed excluding presence of hypervascular focal lesions in the liver and showing patency of the portal vein (PV). The CT scan also showed signs of portal hypertension such as tense ascites, presence of splenogastrorenal shunt, large esophageal and perigastric varices and patency of the umbilical vein. A large volume paracentesis was performed at admission with removal of almost 15 liters of clear ascites. No evidence of infection was found in the fluid (white blood cell count, Gram stain, and culture were negative).

The patient was hemodynamically stable with no need of inotropic drugs. Child-Pugh score was B-8 with a Meld score of 10. She had mild coagulopathy (international normalized ratio 1.4, platelet count 56 000/μL), but with good liver function tests (LFTs) (total bilirubin 1.29 mg/dL, albumin 3.4 g/dL). A transthoracic echocardiography showed no evidence of valvular heart disease, and the diameters of the cardiac chambers were within normal limits. Systolic and diastolic functions of the left ventricle were in the normal range. Estimated pulmonary vein pressure was 35 mmHg.

As a result, we decided to perform a TIPSS procedure in order to treat portal hypertension. The procedure was carried out in an angiographic suite under monitored anesthesia care. The right femoral artery was prepared, and an angiogram revealed patency of the superior mesenteric artery. In the venous phase the superior mesenteric and main portal vein were visualized. After sonographic guidance, the RIJV was cannulated, and under fluoroscopic guidance the metallic flexible guidewire was advanced and seen to curve to the left, until reaching the right atrium (Figure 1). A 5 French (F) pig tail catheter was advanced in the right atrium and a venogram revealed the absence of the right superior vena cava (RSVC) (black arrow) and presence of contrast dye in the left superior vena cava (LSVC) that is draining in the coronary sinus (white arrow).

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Figure 1 Digital venogram showing the curve of the metallic flexible guidewire from the left to the right atrium.

Figure 2 Digital venogram, performed with a 5F pigtail catheter, showing the absence of the right superior vena cava (RSVC) (black arrow) and presence of contrast dye in the left superior vena cava (LSVC) that is draining in the coronary sinus (white arrow).
and laboratory tests every 3 mo. The last US exam was carried out 19 mo post-TIPSS creation showing patency of the TIPSS and absence of ascites. Currently the patient is doing well, her Child-Pugh score is B-7, with stable LFTx and no signs of hepatic decompensation.

**DISCUSSION**

Absence of the right SVC with persistent left SVC in viscero-atrial situs solitus is a rare congenital cardiovascular malformation and one of the most common rare thoracic vein anomalies. It is found in < 1% of the general population and in up to 10% of patients with congenital heart disease[8,9]. The first case was described in 1862 by Halbertsma[10].

This anomaly is usually diagnosed accidentally, for example, during anesthesiological procedures, such as central vein placement[7] or, more frequently, during diagnostic and therapeutic cardiovascular procedures, such as right heart catheterization or pacemaker placement[11,12]. Moreover, it may also emerge during cardiac procedures in pediatric patients (foramen ovale or atrial septal defect transcatheter closure).

Nowadays, TIPSS is considered to be a valid procedure for the treatment of complications of portal hypertension such as refractory ascites and variceal bleeding. The left internal jugular approach has been reported in previous studies[9]. To our knowledge, there are a few cases reported in the literature of successful TIPSS placement through the LSVC approach[13], some of them in patients with a diagnosis of “situs inversus totalis”[11,13]. Interventional radiologists should be aware of this rare, but possible, vascular anatomy, and understand that it may not represent an absolute contraindication for TIPSS insertion.

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