TIPS Creation in a Patient with Situs Inversus Totalis

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Abstract

We describe the successful creation of a transjugular intrahepatic portosystemic shunt (TIPS) in a patient with complete situs inversus using a simple modification of the standard TIPS technique.

Key words: Congenital anomalies—Situs inversus—Left superior vena cava—Shunt, portosystemic

The right internal jugular vein (IJV) is recommended as a standard access for the transjugular intrahepatic portosystemic shunt (TIPS) procedure because there is a lower risk of mediastinal injury than when using the left IJV [1]. The left side is used as an alternative approach in patients in whom the right IJV can not be punctured or in whom portal vein access is unsatisfactory or unsuccessful via the right IJV due to an acute angle between the right hepatic vein and the inferior vena cava [1]. We punctured the left IJV and created a TIPS in a patient with anatomic anomalies of the superior vena cava (SVC) and visceralia: he had a left SVC associated with complete situs viscerum inversus (normal position of the SVC in this anomaly). To the best of our knowledge this is the first report which describes a successful TIPS procedure in a patient with situs viscerum inversus totalis.

Case Report

A 74-year-old man with portal hypertension that had manifested by variceal hemorrhage despite repeated endoscopic sclerotherapy was transferred to our hospital for TIPS creation. Portal hypertension was secondary to end-stage liver disease of cryptogenic etiology. The patient was class A according to the Child-Pugh classification. Endoscopy of the upper gastrointestinal tract revealed varices grade IV. Complete situs viscerum inversus had been known since childhood. US and CT investigations confirmed situs viscerum inversus and revealed mild ascites, splenomegaly, and cirrhotic changes of the liver parenchyma (Fig. 1A).

The interventional radiologist selected the left IJV for access because both the liver and SVC were on the left—this position of the SVC is normal for this anatomic setting (complete situs inversus). TIPS creation was accomplished between the middle hepatic vein and morphologic right portal vein, located on the left side in this case. This procedure was a mirror image of the usual technique: the needle turned to the left when normally it turns to the right if the steel cannula is in the middle hepatic vein. Direct portography depicted portosystemic collaterals arising from the coronary vein (Fig. 1B). The coronary vein was embolized with n-butyl 2-cyanoacrylate (Histoacryl, B. Braun, Melsungen, Germany). The shunt was created using a homemade 12 mm × 40 mm PTFE stent-graft similar to the stent-graft used by Nishimine et al. [2], which was supported by a 12 mm × 60 mm zig-zag stent (Ella stent, Ella-CS, Hradec Králové, Czech Republic) (Fig. 1C). The portosystemic pressure gradient decreased from 14 mmHg to 3 mmHg and the pressure in the portal vein dropped from 24 mmHg to 15 mmHg. The procedure was performed through a 14 Fr sheath (Cook Europe, Bjaeverskov, Denmark).

US follow-up 5 days after the procedure revealed a patent shunt with adequate velocities—maximum velocity in the portal vein was 45 cm/sec, maximum velocity in the shunt was 100 cm/sec—and the patient was discharged. The patient died with a patent shunt 1 month after the TIPS procedure from heart failure.

Discussion

The IJV, SVC and right atrium create a straight channel for manipulation with a stiff cannula in the TIPS procedure. Any congenital anomaly of these veins can cause technical problems.

Congenital anomalies of the SVC are rare, the overall incidence in the general population being 0.3–0.5% [3]. The most common anomaly is left SVC, which is the most frequent anomaly of the thoracic veins [4]. Two types of left SVC have been described. The first type is associated with a normal location of the visceral organs (situs solitus) and the left SVC enters the heart via the coronary sinus. The second is associated with complete situs viscerum inversus. In this type the left SVC enters directly into the morphologic right atrium, which is located on the left side.

We found two cases of SVC anomalies in our group of 400 patients with TIPS, a frequency of 0.4%. One case was left SVC associated with situs inversus totalis and second was persistent left SVC which entered the right atrium via the coronary sinus. The second case has already been described by Bahramipour et al. [5].

Situs inversus is abnormal determination of the visceral organs. Two types of situs inversus can be recognized: complete and partial. Complete situs inversus is defined as a mirror image transposition of both the thoracic and abdominal viscera. In this congenital anomaly the heart lies on the right side of the thorax and the cardiac chambers are reversed. The left lung has three lobes and the right lung has two. The stomach, the spleen, the descending and sigmoid colon are located on the right, while the liver, the ascending colon and the cecum are on the left side. All asymmetric blood vessels are in the reverse position. Total situs inversus is a rare anomaly with an incidence of 0.005% [6]. There is no sex predilection and this abnormality alone is associated with normal life expectancy, intelligence and fertility. The prognosis depends upon
associated congenital malformations, as bronchiectasis is found in 25%, congenital heart disease in 8%, and Kartagener syndrome (situs inversus, bronchiectasis, nasal polyposis or sinusitis) in 10% of patients with situs inversus [7]. The recognition of situs inversus is particularly important when affected persons require surgical or an interventional procedure.

Increasing use of central venous catheterization, implantation of pacemaker leads, catheterization in cardiac examination and endovascular procedures such as TIPS have contributed to more frequent detection of situs inversus totalis during the life of a patient. This anomaly is relatively rare, but an operator must be able to modify the TIPS procedure in its presence. The diagnosis can be anticipated by signs on the chest radiograph, such as a heart shadow and gastric bubble on the right side, accessory interlobium and liver on the left.

The primary use of the left IJV approach was chosen in this case because we anticipated better catheterization of the hepatic vein than by using the right IJV [9]. In such cases we recommend creating a TIPS using a technique which is a mirror image of the usual TIPS procedure.

The right IJV vein approach is usually preferred for the following reasons: (1) there is no lymphatic duct on the right side; (2) the apex of the right lung is lower in most cases than that of the left lung; and (3) the right IJV continues in a straight line with the SVC down to the inferior vena cava while the course from the left side through the left brachiocephalic vein to the SVC is angled, which may cause thoracic pain from stretching of the mediastinal vessels with the stiff steel cannula. Hausegger et al. found [1] the left-sided approach helpful in patients with a nearly 90° angle between the inferior vena cava and the right hepatic vein and in patients with a

Fig. 1. A–C. A 74-year-old man with complete situs viscerum inversus. A Postcontrast CT scan of the abdomen demonstrates situs inversus (the liver and the portal vein are on the left side, the spleen and the stomach are on the right side) and ascites. B Initial transjugular portography shows a patent portal vein and portosystemic varices arising from the coronary vein, which is on the right side. C Final portal venogram after embolization of varices by acrylic glue and stent-graft placement.