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CASE REPORT

A caval homograft for Budd-Chiari syndrome due to inferior vena cava obstruction

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Abstract

Transjugular intrahepatic portosystemic shunt (TIPS) is the standard treatment of Budd-Chiari syndrome (BCS) non responsive to medical therapy. However, patients with inferior vena cava (IVC) obstruction proximal to the atrium do not benefit from TIPS and a surgical approach is mandatory. We report the case of BCS due to intrapericardial IVC obstruction. We describe a novel surgical approach using a fresh caval homograft. An attempt to balloon dilatation of the IVC obstruction was complicated by right atrial disruption with tamponade and ventricular fibrillation. Lately, the patient successfully underwent a reconstruction of the cavo-atrial continuity by the interposition of a fresh caval homograft, a novel surgical approach never described before for BCS. Further follow-up revealed progressive reduction and resolution of ascites, and overall clinical improvement. IVC obstruction near to the atrium can be surgically approached with a new technique consisting in inferior vena cava resection and replacement with a caval homograft.

Key words: Budd-Chiari syndrome; Inferior vena cava; Occlusion; Surgery; Liver transplantation

Core tip: We describe a novel surgical approach using a fresh caval homograft for inferior vena cava (IVC) obstruction proximal to the atrium. An attempt to balloon dilatation of the IVC obstruction was complicated by right atrial disruption with tamponade and ventricular fibrillation. Lately, the patient successfully underwent a reconstruction of the cavo-atrial continuity by the interposition of a fresh caval homograft, a novel surgical approach never described before for Budd-Chiari syndrome. Further follow-up revealed progressive reduction and resolution of ascites, and overall clinical improvement.

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INTRODUCTION

The management of Budd-Chiari syndrome (BCS) non

responsive to medical therapy relies on the possibility of further interventional [angioplasty/stenting or transjugular intrahepatic portosystemic shunt (TIPS)] or surgical (shunts or liver transplantation) approaches[1,2]. However, apart from liver transplantation, surgical approach is performed in a minority of patients[3]. This is probably the consequence of the absence of a well established surgical strategy for BCS, the huge difference in the outcome reported after surgery and the good response homogeneously reported after interventional approaches[4-13]. In fact, TIPS has recently become the standard treatment of BCS non responsive to medical therapy[3]. However, a subgroup of patients with BCS, namely those with inferior vena cava (IVC) obstruction proximal to the atrium, do not benefit from TIPS and a surgical approach is mandatory. The combination of side-to-side portacaval shunt (SSPCS) and cavoatrial shunt seems to be the most reliable surgical technique according to a single centre experience[6].

Herein, we report the case of a patient with intrapericardial inferior vena cava obstruction successfully treated with the reconstruction of the cavo-atrial continuity by the interposition of a fresh caval homograft.

CASE REPORT

A 47 year old man was referred to our centre for ascites and radiological evidence of IVC obstruction.

Due to a Ⅲ degree congenital atrio-ventricular block, a bicameral pacemaker (PM) with epicardial leads had been positioned at the age of 2 years old. Later on, many endocardial leads were implanted and, when he was referred to our centre for ascites, he had 5 catheters in place: two inactive right ventricle (RV) leads trough the left subclavian vein (one since 1980 and one since 1997) and three active leads trough the right subclavian vein (a bipolar atrial catheter since 2010, a RV bipolar catheter since 2010 and a monopolar catheter in the coronary sinus for left ventricle pacing; the 3 active catheters were linked to a ABIV PM).

At admission, examination revealed ascites and leg swelling. Biochemistry revealed hemoglobin 11.1 g/dL, platelets 173000/mm^3, albumin 4.2 g/dL, normal aspartate aminotransferase/alanine aminotransferase, only slightly increased alkaline phosphatase and gamma-glutamyltransferase, bilirubin 1.8 mg/dL, international normalised ratio 1.3. Splenomegaly was evident clinically and at ultrasound (14 cm). A contrast-enhanced computed tomography confirmed a complete IVC occlusion at the level of the connection with the right atrium. At that level, one of the inactive leads formed a loop with presence of evident ingrowth of fibrotic tissue. The angiography confirmed the occlusion of the IVC with an extension of 2 cm.

In November 2011, in the cardiac hybrid suite, an attempt to balloon dilatation of the IVC obstruction was performed but was complicated by right atrial disruption with tamponade and ventricular fibrillation. An emergency sternotomy was performed and the atrial tear was repaired with a running suture without the institution of the extracorporeal circulation. The patient was then admitted to the intensive care unit and slowly recovered despite progressive worsening of ascites and leg swelling.

In December 2011, an elective surgical treatment was planned. A jugulo-pubic incision with median sternotomy was performed. Hepatic hilus was prepared and through a phrenic incision the junction between the inferior vena cava and the right atrium was thoroughly isolated. The extracorporeal circulation was instituted between aorta, superior vena cava and right femoral vein. On beating heart, the right atrial wall was opened in the site of the previous tear and all the leads were removed. The obstruction of the vena cava was due to the excess of fibrous tissue around one of the old inactive electrodes which migrated deeply into the vein. All the scarred tissue was removed and the hepatic vena cava was opened free with the supra-hepatic veins widely pervious. Then the liver and the right atrium were connected with a segment of vena cava harvested from a cadaveric multi-organ donor and preserved in electrolytic solution. The extracorporeal circulation was interrupted and the inferior caval flow restored. Three new epicardial electro-catheters were implanted, respectively, one on the left ventricle, one on the RV and one in the right atrium. Few days after surgery ascites and swelling started to resolve and the patient was discharged on day 12 with low dose of diuretics that were subsequently stopped.

Histological examination of the liver biopsy performed during surgery confirmed a BCS picture and revealed severe centro-central bridge and sinusoidal fibrosis.

At the programmed examinations, respectively three, six and thirteen months after surgery, the patient was well, with no signs of inferior venous congestion and normal liver biochemistry. Moreover doppler echography revealed a normal flow from IVC and right atrium.

DISCUSSION

BCS is a rare and serious disease with a generally worsening outcome without intervention. Although definition of response to treatment is a debated topic, when medical treatment alone is not sufficient to prevent progression, further treatments are needed[1,13].

It is widely accepted that the management of BCS should follow a step by step strategy. In fact, medical therapy is the first-line treatment, angioplasty/stenting or TIPS the further step and liver transplantation (LTx) the last chance[1,2].

Apart from LTx, surgical treatment is generally not contemplated in the BCS management by recent guidelines[1,2]. In fact, the most used treatment for BCS non responsive to medical therapy is TIPS, LTx is used as a rescue therapy, while surgical treatments are limited to a strict minority of patients[3].

TIPS is surely the mostly used treatment for BCS when medical therapy fails[3]. In early experiences, TIPS has proved effective as BCS treatment, also in the technically difficult case of extension of thrombosis to the por-

tal vein tree[10,11]. Recently, a multi-centre study provides long-term data on TIPS treatment for 147 BCS with a 10 year survival of 69%[12]. A less used radiological treatment is angioplasty/stenting[8,9].

Surgical shunts for BCS can be very successful, but have been associated with rapid decompensation and in-hospital mortality can be high (about 25%), primarily due to the patients' poor general condition[4-8]. In the past years, a surgical approach has been traditionally considered the first choice. In some series, an excellent outcome with long-term follow-up has been reported (95% survival, 3 to 28-year follow-up)[6]. However, in this series, the SSPCS was used, an approach that cannot be used when there is IVC thrombosis or significant compression. In the same series, the mortality rates of patients with IVC involvement were very high after traditional surgery (mesoatrial shunts) and better results were described with another technique (SSPCS + cavoatrial shunt) in 18 patients, all survived after a follow-up of 5-25 years[6]. However, outcome after surgical portosystemic shunt is variable and worse results were reported by others[4,5,7,8]. In most of the above series, patients with liver failure were not considered for surgery but for liver transplantation[4-8].

LTx is the last chance for BCS syndrome non responsive to either medical therapy and re-canalization/decompression. A multi-centre study reported a 10 year survival of 68%[14].

Patients with BCS due to a IVC obstruction near to the atrium constitute a difficult to treat subgroup of BCS patients, whose management can benefit from endovascular dilatation/stenting or surgical treatment.

Because of obvious reasons, TIPS does not by-pass IVC obstruction, resulting ineffective. Although some study describe the possibility of endovascular management as safe and with good long-term patency, data are scanty and need confirmation on a larger scale[15].

The patient we describe underwent angiographic IVC obstruction balloon dilatation that was complicated with atrial laceration and pericardial tamponade , requiring immediate sternotomy, pericardiotomy, internal cardiac massage, pericardial blood drainage and atrial suture. Surgical approach to BCS due to a IVC obstruction near to the atrium relies on the possibility of surgical shunts or LTx . Although the long-term results of LTx for BCS are good, we believe that LTx should be the last change, since surgical correction avoid successive immunosuppression and the possibility of LTx complications[14].

The widest experience with a surgical shunt approach for BCS due to a IVC obstruction is that recently published, and the best experience was reported after a combination of SSPCS and cavo-atrial shunt: 18 patients, all survived after a follow-up of 5-25 years[6]. Other surgical techniques reported in that and other series either had unacceptable or non reported outcomes[4,5,7,8].

The case here reported had an excellent outcome after the replacement of the obstructed segment of the IVC with a caval homograft. This technique requires a strict collaboration with the hepatic and the cardiac team, with experience both in cardiac and hepatic transplantation, and the aid of the extracorporeal circulation. The key of success is to obtain a clear surgical field, with all the blood drained from the right atrium and from the liver, allowing a perfect control of the anastomotic site. Moreover the use of a fresh homograft facilitates immensely the surgical performance.

Acquired inferior vena cava obstruction near to the atrium can be surgically approached with a new technique consisting in IVC resection and replacement with a caval homograft.

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