Endovascular Management of Budd Chiari Syndrome - A Case Report

Abstract

Budd Chiari syndrome (BCS) Is a diverse group of diseases which present with clinical features of hepatic venous obstruction or inferior vena cava obstruction. This syndrome is usually treated with medical management. We present a case of a young male patient who was diagnosed with BCS. He was found to have a total occlusion of retro hepatic inferior vena cava. He was treated with endo vascular methods - venoplasty and stenting. This case report highlights the presentation and diagnosis of this relatively rare syndrome. By treating the patient with endovascular methods we were able to give a satisfactory outcome which led to early recovery.

Introduction:

Budd Chiari syndrome (BCS) denotes a heterogeneous group of diseases characterised by hepatic venous outflow obstruction at the level of hepatic veins or inferior vena cava. This syndrome can be fulminant, acute, chronic, or asymptomatic. It occurs in 1 out of 100,000 individuals and is more common in females. Traditional management of BCS involves thrombolysis or surgical portosystemic shunt creation. This case report showcases a successful management of BCS by endovascular treatment.

Case Report:

A 28 yr old male patient presented to our outpatient department with complaints of pain and swelling in both lower limbs since 6 months. He was a previously healthy man with no significant medical or family history. The physical examination was unremarkable except for bilateral pitting pedal edema with a few dilated veins over both lower limbs. There was no evidence of arterial insufficiency. His abdomen was soft, non tender with no organomegaly.

The blood investigations which included a complete blood count, renal function tests and liver function tests were normal. He had a normal albumin level of 4.2 g/dl. The bleeding and clotting parameters were within normal range. The INR on admission was 1.1. Ultrasound scan of the abdomen was done as a preliminary diagnostic imaging. This showed mild hepatomegaly with no ascites. A CT Venogram of the abdomen revealed a total occlusion of retrohepatic inferior vena cava.

In view of the young age of the patient we decided to attempt an endovascular treatment. After gaining access through the right femoral vein with 10F catheter and right jugular vein access with 6F catheter a selective venacavogram was performed. This demonstrated a total thrombotic occlusion of the inferior venacava in the retrohepatic segment for a length of about 40 mm. The occlusion was carefully crossed with a stiff tip wire. Balloon dilatation of the occluded segment was done with 12x4 and 16x4 cm balloons. To keep the venacava patent 24x45 mm Wall self expanding stent was placed. The stent was post dilated with 22x4 cm balloon. At the end of the procedure, the completion venogram showed a patent stent with no residual occlusion.

Patient was discharged home the following day with antiplatelets and he is on regular follow up with no further complications related to the procedure.

Discussion:

Budd Chiari Syndrome has been studied with interest since many years. The etiology of BCS could be primary or secondary. Primary BCS is mainly due to intrinsic intraluminal thrombosis or webs in the venacava. It can be secondarily caused by extraluminal compression (by abscess or tumours) or intraluminal invasion by tumours or parasites. The most frequent cause of BCS in Western countries is thrombotic

occlusion in a hyper coagulable state. The common clinical presentations include ascites, abdominal pain, hepatomegaly, dilated veins on the trunk, leg edema and jaundice.

The traditional management of BCS involves thrombolysis or surgical porto systemic shunt creation. Both these approaches have significant morbidity and procedure related complications. An increasing number of successful reports of BCS therapy have involved endovascular techniques, including angioplasty and stent placement. These techniques are especially effective in cases of hepatic or IVC thrombosis or webs. In this case report we treated the patient with a minimally invasive technique. The occlusive segment of the vena cava was made patent with the placement of stent, thereby restoring flow. The lower limb oedema which was secondary to the obstructed venous drainage was relieved after the stent placement. This approach could be a first line treatment with appropriate patient selection.

References:

- Budd-Chiari Syndrome: Hepatic Venous Web Outflow Obstruction Treated by Percutaneous Placement of Hepatic Vein Stent Alireza Bozorgmanesh, D. Arul Selvam, James G. Caridi . Semin Intervent Radiol. 2007 March; 24(1): 100–105.
- Bogin V, Marcos A, Shaw-Stiffel T. Budd-Chiari syndrome: in evolution. Eur J Gastroenterol Hepatol. 2005;17:33–35.
- Lee B B, Villavicencio L, Kim Y W, et al. Primary Budd-Chiari syndrome: outcome of endovascular management for suprahepatic venous obstruction. J Vasc Surg. 2006;43:101–108.

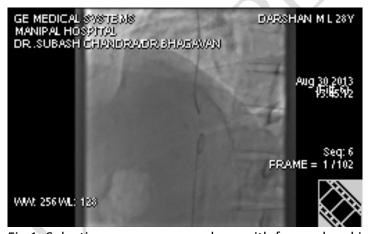


Fig 1: Selective venacavogram done with femoral and jugular access

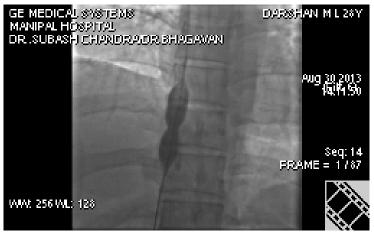


Fig 2: Balloon dilatation done with 12x4 and 16x4 cm balloons

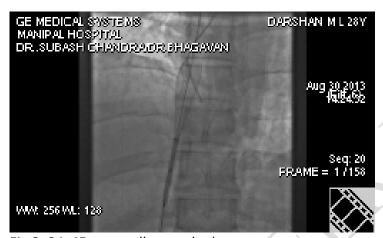


Fig 3: 24x45 mm wall stent deployment

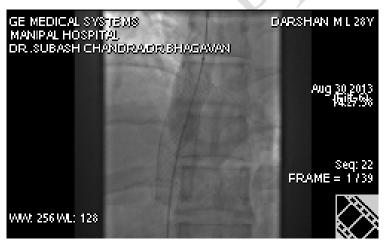


Fig 4: Post stent dilatation with 22x4 cm balloon

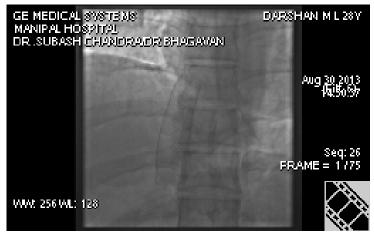


Fig 5: Final result