Endovascular Management of Budd Chiari Syndrome - A Case Report

ABSTRACT:

Background:
Budd Chiari Syndrome denotes a group of rare diseases which have been studied with considerable interest by clinicians across the world. The clinical presentation and investigation results vary in most cases making the diagnosis a challenge. Although the treatment protocols are published in literature, much of it comes from expert opinions rather than research-based findings. The use of medical therapy, thrombolysis and interventional procedures are well known. However, the order of preference while choosing the treatment option depends on each case and must be considered depending on local expertise.
Case summary:
We present a case of a young male patient with no significant medical history who presented with lower limb oedema and pain. The laboratory findings were within normal range and the ultrasound scan revealed mild hepatomegaly. On further imaging we found a retrohepatic vena caval occlusion which was amenable to endovascular intervention. This patient was treated with angioplasty and stenting with satisfactory clinical outcome. The decision to proceed with an endovascular first approach is highlighted in our case report.
Conclusion:
The traditional treatment strategies for BCS include medical therapy, thrombolysis or surgical portosystemic shunt creations which involve significant morbidity and procedure related complications. It is worthwhile to select patients with amenable lesions to undergo endovascular treatment in the initial stages of management. This will lead to good clinical outcomes and prevent long term liver damage. This approach has been highlighted in our case report and we recommend its use with the availability of local expertise.

Key words: Budd Chiari Syndrome, inferior venacava, IVC, venoplasty, endovascular

INTRODUCTION :

Budd Chiari syndrome (BCS) is a rare group of diseases characterized 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. We present a case report of a young male patient who presented with innocuous complaints of lower limb swelling and pain with no abdominal symptoms. Although his blood investigations were within normal range, the ultrasound scan showed mild hepatomegaly. Further imaging revealed a total occlusion of retrohepatic inferior venacava (IVC) and the patient was treated with venoplasty and stenting. Traditional management of BCS involves thrombolysis or surgical portosystemic shunt creation. This case report showcases a successful management of BCS by endovascular first treatment. We believe that the use of angioplasty/stenting should be considered early on during the treatment planning along with the use of medical therapy. This will lead to a successful initial outcome and avoid long term liver damage.

PRESENTATION OF CASE:

A 28-year-old male patient presented to our outpatient department with complaints of pain and swelling in both lower limbs for 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 international normalized ratio (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 Computed Tomography Venogram of the abdomen revealed a total occlusion of retrohepatic inferior vena cava.

In view of the young age of the patient and the lesion characteristics 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.

Due to the rarity of BCS, the management protocols have evolved based on expert opinion rather than evidence-based experiences. In 2009 DeLeve et al proposed a step wise management which suggests moving forward when no response to medical therapy appears[1]. Accordingly, the first line treatment would be the medical therapy (anticoagulation, treatment of underlying disease, symptomatic therapy of portal hypertension complications), angioplasty/stenting the second-line (in patients with short-length stenoses not responding to medical therapy), Transjugular intrahepatic portosystemic shunt (TIPS) the next step (in patients not responding to medical therapy and in case of no response to, or stenoses unsuitable for, angioplasty/ stenting) and liver transplantation (LT) the last chance when TIPS is not effective.
This management strategy for BCS was challenged by Mancuso in 2014 who proposed a new algorithm[2]. In this argument the invasive treatments such as TIPS and angioplasty should be considered before no response to medical therapy appears. This could perhaps decrease hepatic fibrosis development, disease progression and finally improve outcome.

An increasing number of successful reports of BCS therapy have involved endovascular techniques, including angioplasty and stent placement[3-6]. These techniques are especially effective in cases of hepatic or IVC thrombosis or webs. When this approach is used for short length stenoses a good medium-term outcome is reported in some experience[10-12].

In this case report, the patient had symptoms of IVC obstruction in the absence of any previously known pro thrombotic conditions. After starting the patient on medical therapy, we decided to treat the lesion with endovascular techniques. 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. The successful treatment in this case was due to the concurrent use of endovascular treatment along with medical therapy.

CONCLUSION:

We believe that an endovascular-first approach in the initial management of BCS should be considered for all cases with amenable lesions. The availability of local expertise is essential for this approach to be successful. When applied judiciously and on a case-by-case assessment, the use of endovascular therapy in BCS will lead to a good initial outcome and prevent long term liver damage.

Consent Disclaimer:

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

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