Case Report

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Balloon cavoplasty in membranous obstruction of vena cava

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ABSTRACT

A hepatic venous outflow tract obstruction at any level is considered as Budd Chiari syndrome (BCS). Primary BCS is usually due to a congenital membrane causing the obstruction; referred to as membranous obstruction of vena Cava (MOVC). In the past MOVC was predominantly treated through surgery, percutaneous transluminal balloon angioplasty (PTBA) is an alternative and effective form of treatment. Case scenario of a 32-year-old gentleman presented himself with complaints of hematemesis for one year and hematochezia for three months. Hepatomegaly was noted. An ultrasonogram revealed a dilated IVC, till its hepatic portion, and also the Hepatic Veins. There was a fibrous membrane like structure of about 5 mm thickness that was obstructing the IVC flow. Patient was taken up for venogram for conformation of diagnosis and intervention. 6F NIH catheter was introduced from Femoral vein, an injection into the IVC showed contrast not flowing into right atrium and there was a membranous obstruction for the forward flow. A Brockenbrough's trans-septal needle with Mullins Sheath was used to puncture the membrane and right atrium was entered. A 22 mm Inouye mitral valvuloplasty balloon (Single Balloon, Toray Medical, Tokyo) was taken and positioned at the membrane and inflated and deflated several times with incremental pressures till the waist of the balloon disappeared. Conclusion membranous obstruction of vena cava (MOVC) is a common cause of primary Budd Chiari syndrome. Percutaneous transluminal angioplasty using Inoue or mansfield balloon with or without stenting is a safe and effective treatment option.

Keywords: Balloon Cavoplasty, BCS, MOVC, PTBA

INTRODUCTION

Membranous Obstruction of Vena Cava (MOVC) is an unusual but potentially treatable cause of Budd-Chiari Syndrome(BCS). These membranes are thought to be a congenital abnormality but they are more frequently acquired through a thrombotic process secondary to myelodysplastic disorders or hepatocellular carcinoma.¹

BCS is caused by hepatic venous outflow tract obstruction at any level, provided that the obstruction is not caused by cardiac, pericardial or veno-occlusive disease. In Asian countries, it is predominantly seen in

males with mean age of diagnosis being 45 yrs. Also, pure Inferior Vena Cava (IVC) or combined IVC and Hepatic Vein obstruction is more common in Asian population. In Western countries BCS is found mostly in women and its predominant cause is hepatic vein obstruction.² BCS is considered as primary when the obstruction is predominantly due to thrombosis or phlebitis and secondary when veins are being compressed due to a lesion.

In the past MOVC was predominantly treated through surgery Percutaneous Transluminal Balloon Angioplasty(PTBA) is an alternative and effective form of treatment. Here we present a case of a 32y gentleman who was diagnosed to be having primary BCS due to IVC membrane and underwent PTBA.^{3,4}

CASE REPORT

A 32-year-old gentleman presented himself with the complaints of hematemesis for one year and hematochezia for three months. He was not an alcoholic. He has been smoking ten cigarettes per day for the past fifteen years. On examination, there were prominent veins over the abdomen and it was distended, shifting dullness was present. Hepatomegaly was noted. An Ultrasonogram revealed a dilated IVC, till its hepatic portion, and also the Hepatic Veins were dilated. There was a fibrous membrane, of about 5 mm thickness, that was obstructing the forward flow. There was no forward flow in the hepatic portion, but it was present in the lower portion of Inferior Vena Cava. The other organs of abdomen were unremarkable. This picture suggested of a primary BCS.

Patient was taken up for venogram for conformation of diagnosis and possible intervention. Right femoral and right internal jugular vein access was taken and 6F sheaths were introduced. 6F NIH catheter was introduced from Femoral vein, an injection into the IVC showed no contrast flow into right atrium and there was a membranous obstruction for the forward flow. To further delineate the membrane, two 6F NIH catheters were introduced one from right femoral and the other from right internal jugular vein. Simultaneous contrast injection revealed a thin membrane that was just cephalad to the entry of Hepatic Vein.



figure 1: RAO cranial view, simultaneous injections using 6F NIH catheters in internal jugular and femoral vein. A membranous obstruction can be seen clearly.

A Brockenbrough's trans-septal needle with Mullins Sheath was used to puncture the puncture membrane and right atrium was entered. The pressure in right atrium was measured. After dilating the puncture with Mullins dilator, a Coiled tip wire was positioned in right atrium. A 22 mm Inoue Mitral valvuloplasty Balloon (Single

Balloon, Toray Medical, Tokyo) was taken and positioned at the membrane and inflated and deflated several times with incremental pressures till the waist of the balloon disappeared.

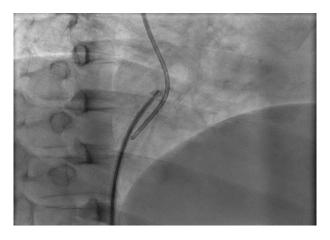


Figure 2: Puncturing the membrane and RA is entered using brockenbrough's trans septal puncture needle.

The balloon was removed and the IVC venogram was done which revealed an unobstructed flow to the right atrium and the pressure gradient between the IVC segment and right atrium also fell. Patient tolerated the procedure well, his symptoms and signs improved over the next two days. He was put on anticoagulation with Warfarin for 3 months (INR at 2-2.5), after over lapping with Heparin, and discharged. One year from the procedure patient is doing well with no recurrence of symptoms or signs.

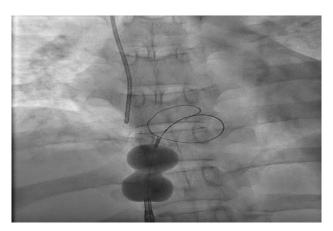


Figure 3: Dilatation of the membrane using 22 mm Inoue balloon.

DISCUSSION

BCS can have a varied presentation. Fulminant liver cell failure is characterized by acute liver injury, elevation of transaminases, encephalopathy, deterioration of prothrombin time occurring within eight weeks of development of jaundice. In the acute form, clinical manifestations and deterioration of liver functions occurs

in several weeks. In subacute form, there is an insidious onset and clinical features develop within three months. In chronic form, patient usually presents with complications secondary to cirrhosis of liver.

50% of BCS cases are caused due to an underlying myeloproliferative disorder chronic with accompanying hyper coagulable state. JAK2 Tyrosine kinase mutation was found in 26 to 59% of patients diagnosed with BCS.5 JAK2 mutation is seen in patients of polycythaemia vera and also 50% of patients of thrombocytosis or chronic essential idiopathic myelofibrosis.6 Malignancies account for approximately 10% of cases of BCS, most common being Hepatocellular carcinoma. Infections of the liver (cysts, abscesses) and benign lesions (Adenoma, Cystadenoma) constitute another 10% of cases. Other causes include Oral contraceptive usage and pregnancy, hyper coagulable states (Factor V Leiden mutation, Protein C and S deficiency, antithrombin deficiency etc.) and Membranous Obstruction Vena Caval (MOVC) is the cause in approximately one third of all cases, it is much more common in South Africa, India and Asia. In close to 20% of patients the cause is not found and they are considered idiopathic.

Due to its varied presentation, as outlined above, BCS must be considered in the differential diagnosis of patients, with predisposing factors, presenting with acute or chronic liver disease. Diagnosis is usually done by doppler ultrasonography. CT or MRI scan can confirm the diagnosis and aide in treatment planning. Venography is the gold standard for diagnosis and is usually done when non-invasive tests are inconclusive. Venography is also vital for directing therapy. Patients must also undergo an evaluation for predisposing conditions like prothrombotic states.

The prognosis of patients who are symptomatic is bad, as 90% of people die within a period of three years. With an appropriate intervention, the prognosis is good. The one, two and five-year survival rates are 87, 82 and 74 % respectively with an appropriate intervention. The therapeutic options available are anticoagulation and diuretics, thrombolysis, percutaneous transluminal angioplasty, trans jugular intrahepatic portosystemic shunting, surgical portosystemic shunting and orthotropic liver transplant.

Features like older age at presentation, severe liver failure with refractory ascites, involvement of portal veins are associated with poor prognosis. ¹⁰ Anticoagulation can be considered in patients where risk factor is deemed to be permanent or when a thrombophilia is detected. However prospective randomized control trials are lacking in the case of BCS. Thrombolysis may be considered when the clot that is causing the BCS is recent, i.e. within three to four weeks old. There is scanty evidence through case series, of patients where insitu thrombolysis was done

successfully.¹¹ Thrombolysis is not an option in chronic cases

Surgical portosystemic shunting is another modality for decompression of liver, but the periprocedural mortality is very high approaching 23%.12 Percutaneous trans luminal angioplasty of IVC/Hepatic Veins with or without stenting decompresses the liver and allow for immediate improvement in signs, symptoms and liver functions. The periprocedural mortality in transluminal approach is very minimal. Balloon angioplasty is an excellent option when the abnormality is focal. Balloon dilatation can be done in Membranous obstruction in IVC, with high periprocedural success rate. 13 Dilatation of the membrane or segmental stenosis is done using Inoue balloon or Mansfield balloon. The major drawback of balloon dilatation is the recurrence of stenosis. This can be avoided by deploying a metallic stent. There has been recent evidence that stent implantation can increase the patency rates.¹⁴ Stent implantations has also been shown to improve liver function tests and regress portal hypertention.¹⁵ The decision to stent depends of the type of the lesion. Long segment, multiple segments of obstruction, restenosis after balloon dilatation, nodular compression of IVC all do better with stenting.16 Hepatic vein intervention with angioplasty or stenting has also been done with success and the severe periprocedural complications appear to be minimal.¹⁷ Liver transplant appears to be the only option for patients who fail the minimally invasive options or are in decompensated cirrhosis.

CONCLUSION

Balloon angioplasty must be considered in cases of MOVC where a focal obstruction is causing the symptoms. Decision to stent can be individualized; where long lesions, multiple lesions, restenosis and compression of IVC are better tackled with implantation of a stent.

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