Successful Endovascular Stenting of the Inferior Venacava (IVC) in a Patient with Budd-Chiari Syndrome (BCS)

Dr. P. Sampath Kumar, Dr. V. S. R Bhupal, DR. K. C. Karthik Naidu, Dr. G. Pranoy

Abstract: A 60 year Old man an Agricultural labourer by occupation had Distension of Abdomen, Prominent of veins over abdomen & Swelling of both legs with excoriation for the past 8 years was diagnosed as Budd chiari syndrome with IVC Obstruction. MR venogram showed focal short segmental stenosis causing 80-90 % luminal narrowing of IVC at diaphragmatic hiatus region with mild downstream dilatation of IVC which is 33 mm. IVC angiogram was done which showed obstruction at the junction of IVC and right atrium with thickness of membrane around 5mm. IVC SELF EXPANDING WALL STENT (18 x 60mm) was done and IVC flow was restored.

patient symptoms reduced & he is on regular follow up.

Keywords: PT-INR (Prothrombin-International Normalized ratio), ECG (Electrocardiogram), CBD(common Bile duct), IHBD (Intra-Hepatic biliary Dilatation), PCS (pelvicalyceal System), CMD (Cortico Medullary differentiation), IVC (Inferior venacava), SFV (superficial femoral vein), IJV (internal jugular vein), MPA (Multipurpose) LAO (left anterior Oblique), RA (Right Atrium), RV (Right Ventricle), LA (Left Atrium)

1. Introduction

A 60 year Old man an Agricultural labourer by occupation had Distension of Abdomen, Prominent of veins over abdomen & Swelling of both legs with excoriation for the past 8 years was diagnosed as Budd chiari syndrome with IVC Obstruction. MR venogram showed focal short segmental stenosis causing 80-90% luminal narrowing of IVC at diaphragmatic hiatus region with mild downstream dilatation of IVC which is 33 mm. IVC angiogram was done which showed obstruction at the junction of IVC and right atrium with thickness of membrane around 5mm. IVC SELF EXPANDING WALL STENT (18 x 60mm) was done and IVC flow was restored. patient symptoms reduced & he is on regular follow up.

A 60 year Old man Mr. X, An Agricultural labourer presented to the hospital with complaints
1) Distension of Abdomen,
2) Prominent of veins over abdomen,
3) Swelling of both legs with excoriation for the past 8 years.

Routine investigations

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>9.6 g%</td>
</tr>
<tr>
<td>Total count</td>
<td>7500 cells/cumm</td>
</tr>
<tr>
<td>Serum creatinine</td>
<td>1.1 mg/dl</td>
</tr>
<tr>
<td>Serum Sodium</td>
<td>146 meq/L</td>
</tr>
<tr>
<td>Serum Potassium</td>
<td>4.4 meq/L</td>
</tr>
<tr>
<td>PT – INR</td>
<td>1.73</td>
</tr>
<tr>
<td>Viral markers</td>
<td>Non Reactive</td>
</tr>
</tbody>
</table>

LFTs (Liver function tests)

Serum Total bilirubin = 2.5mg%
Direct Bilirubin = 1.0 mg%
Indirect bilirubin = 1.5 mg%
SGOT = 200 IU/L
SGPT = 256 IU/L
Alkaline phosphatase = 125 IU/L
Serum Total proteins = 4.8 g%
Serum Albumin = 2.8 g%
Serum Globulin = 2.0 g%
A/G ratio = 1.4

Roentogram

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Volume 9 Issue 7, July 2020
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Paper ID: SR20707150842  DOI: 10.21275/SR20707150842
ECG

Ultrasound abdomen (21-08-18)

- **Liver**—measures 14.8 cm. Normal in size with shrunken right lobe & hypertrophied Caudate lobe & coarsed Echopattern
  
  No IHBD. PV = 13.9 mm, CBD – Normal caliber

- **Gallbladder**—Normal

- **Spleen**—Enlarged in size measuring 15.0 cm. shape and echo pattern are normal

- **Kidneys**—Normal in size and echopattern. PCS normal. CMD made out and No calculi

  - **Impression**—Cirrhosis of Liver with Splenomegaly

MRI VENOGRAHM OF IVC:

a) Features of cirrhosis of liver with portal hypertension

b) Showed **focal short segmental stenosis** causing 80-90% luminal narrowing of IVC at diaphragmatic hiatus region with mild downstream dilatation of IVC which is 33 mm.

c) Overall features probably represent –Budd chiari syndrome with cirrhosis of liver
- **2D ECHO**: Dilated IVC & narrowing of IVC before entering into right atrium.
- Good LV function
- No LV RWMA
- NO MR/AR/PR/TR
- Grade I Diastolic dysfunction

IVC angiogram was done which showed obstruction at the junction of IVC and right atrium.

The thickness of the obstruction was assessed by passing catheters from femoral and jugular approach which showed the thickness to be 5 mm.

**Approach through Right SFV** – engaged with MPA1 crossed IVC till the stenotic site. **Approach through Right IJV** engaged with JR 3.5 into the RA.
MPA removed – Introduced Mullen sheath along with septal puncture needle

Confirming position of needle in LAO & left lateral Position at 12 o clock position.
Septal needle is advanced through the stenotic site entering the RA.

Over the septal puncture needle – Mullens sheath was advanced into the RA.
LA wire was passed into the RV – there after Mullens sheath is removed

20 x 20 mm balloon is passed over the LA wire – Inflated to 20 atm
Balloon removed – Over the wire _ IVC SELF EXPANDING WALL STENT (18 x 60mm)

IVC wall Stent (18 x 60mm)
Finally, Contrast was introduced into IVC which showed good flow into the RA.
• Prominent veins over the Abdomen - disappeared with in 1hr of procedure
• The patient began to lose body weight - 1 day after stenting
• Day 2 the patient Ascites and pedal edema slowly disappeared.
IVC Doppler – Post procedure

1) **IVC STENT NOTED INSITU**
2) Rest of the IVC shows normal collapse and unidirectional flow
3) No obvious thrombosis noted within visualized part of IVC

Follow up 2D echo after 1 month – Mainly IVC

On treatment & regular follow up
1) Tab. Warf 2mg/3mg alternate days
2) Tab. Atocor CV 75/10 mg po H/S (Atorvastatin 10mg + Clopidogrel 75mg)

**Budd-Chiari Syndrome (BCS)**
This a rare syndrome occurs in 1/1, 00, 000 in the general population.

Budd Chiari syndrome is a heterogenous group of disorders characterized by Hepatic outflow obstruction at the level of hepatic venules, the large hepatic veins, IVC (or) the Right atrium

Hepatic veno-occlusive disease refers to the obstruction of Hepatic venous outflow at the level of central (or) Sub-lobular hepatic veins (or) both

**Hepatic vein (or) IVC lesions** include
- Membranous (or)
- Stenotic obstruction (or)
- Thrombosis.

**Volume 9 Issue 7, July 2020**

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Paper ID: SR20707150842
DOI: 10.21275/SR20707150842
Thrombosis of the hepatic veins is the most common cause of BCS in Western countries. Whereas Membranous obstruction of the IVC (or) the Hepatic veins is more common in Eastern countries. Patients with hepatic vein occlusion may present with symptoms and Signs of portal hypertension including Ascites, hepatomegaly, splenomegaly and progressive liver dysfunction.

Two underlying causes lead to a manifestation of BCS.
One is Hepatic vein thrombosis, which is often associated with various coagulation defects and disorders.
The other is Membranous obstruction of hepatic veins (or) the IVC which occurs more common in Eastern countries and may be congenital in origin.
As most of the significant symptoms and signs in patients with BCS are Associated with portal hypertension, treatment is usually intended to relieve portal hypertension.

Table 1. Causes of the Budd–Chiari Syndrome.

<table>
<thead>
<tr>
<th>Common causes</th>
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<tbody>
<tr>
<td>Hypercoagulable states</td>
</tr>
<tr>
<td>Inherited</td>
</tr>
<tr>
<td>Antithrombin III deficiency</td>
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<tr>
<td>Protein C deficiency</td>
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<tr>
<td>Protein S deficiency</td>
</tr>
<tr>
<td>Factor V Leiden mutation</td>
</tr>
<tr>
<td>Prothrombin mutation</td>
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<tr>
<td>Acquired</td>
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<tr>
<td>Myeloproliferative disorders</td>
</tr>
<tr>
<td>Paroxysmal nocturnal hemoglobinuria</td>
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<tr>
<td>Antiphospholipid syndrome</td>
</tr>
<tr>
<td>Cancer</td>
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<tr>
<td>Pregnancy</td>
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<tr>
<td>Use of oral contraceptives</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Uncommon causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tumoral invasion</td>
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<tr>
<td>Hepatocellular carcinoma</td>
</tr>
<tr>
<td>Renal-cell carcinoma</td>
</tr>
<tr>
<td>Adrenal carcinoma</td>
</tr>
<tr>
<td>Miscellaneous</td>
</tr>
<tr>
<td>Aspergillosis</td>
</tr>
<tr>
<td>Behçet’s syndrome</td>
</tr>
<tr>
<td>Inferior vena caval webs</td>
</tr>
<tr>
<td>Trauma</td>
</tr>
<tr>
<td>Inflammatory bowel disease</td>
</tr>
<tr>
<td>Dacarbazine therapy</td>
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<tr>
<td>Idiopathic</td>
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</tbody>
</table>

Porto-diversion treatment, such as Side-to-Side portocaval, Mesocaval and Mesoatrial shunts have been used, but these surgical methods are also associated with many serious complications.
Recanalization of occluded major hepatic veins (or) the IVC facilitates hepatic venous outflow drainage
Thus the hepatic congestion is relieved & portal pressure decreases, which is beneficial for recovery of liver function.
Endovascular treatment has become a reliable treatment option for patients with symptomatic BCS.
Restoring hepatic venous outflow is the primary objective when treating BCS. Endovascular treatment is the main therapeutic choice with advances in interventional technology.

A trans-jugular intrahepatic portosystemic shunt (TIPS) is currently the most common intervention for patients with BCS in Western countries because obstruction of the hepatic veins is usually extensive.
Balloon dilatation (or) Stent insertion can be attempted to recanalize the Hepatic venous outflow pathways, when the lumen of a hepatic vein (or) the IVC is partially maintained.
Hepatic venous pressure can Drop 25-50% after successful endovascular stenting in patients with BCS.
The 5-year survival rate in patients with mild disease and preserved liver function can be as high as 100%.
Survival rates for patients with progressing BCS are about 85% at 1 year and 77% at 5 years.
The common complications of endovascular treatment for BCS include inadvertent IVC perforation
The reported overall incidence of complications is <3%, and mortality rate is about 1%.

The incidences of re-stenosis after balloon expansion and stenting of hepatic veins are 10-20% and 10% respectively.

References