IDIOPATHIC MEMBRANOUS IVC OBSTRUCTION - A CASE REPORT

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Abstract:
An Inferior Venacaval (IVC) lumen obstruction, whether it is partial or complete, is not a common phenomenon and it may be caused by the thrombosis or compression which may result from neoplastic or non neoplastic masses. There are cases which are idiopathic, typically referred to as membranous obstructions of the IVC. Here, we are presenting a 13 years old boy with multiple tortuous veins over the abdomen, with ascites, who underwent a successful percutaneous transluminal balloon dilatation of the IVC membrane which was located above the hepatic veins, which resulted in a successful palliation of his symptoms.

Keyword :
Membranous IVC Budd Chiari syndrome

Introduction:
An Idiopathic Obstruction of the Inferior Vena Cava (IOIVC) is common in some parts of the world. A series of such cases was described by the authors in Japanese institutions1,2 where IOIVC was a more common cause of the Budd-Chiari Syndrome (BCS) than the more classical intrahepatic venous obstruction. The IOIVC is also relatively common in the black population of South Africa5,7. In the Indian population, the lesions are generally situated within the hepatic portion of the IVC, below or at the diaphragmatic level(3,4,8). Many have associated obstruction of one or both of the main hepatic veins, a finding which permits separation of the lesions of the IOIVC into eight different categories. The commonest form appears as a thin membrane which runs below an obliterated left hepatic vein, but above a patent right hepatic vein.

Case Report:
A 13 years old boy, presented with multiple dilated tortuous veins over the abdomen, extensively over the central and the lateral abdomen and the back, which extended downwards into both the inguinal regions, with mild abdominal distention. The flow in the dilated tortuous veins appeared to be below upward. He had NYHA class II dyspnoea. An abdominal ultrasonography revealed narrowing of the suprahepatic inferior venacaval lumen with mild ascites. No definite membrane could be visualized. A Computed Tomography (CT) of the abdomen which was done elsewhere,
was not informative and it was suggestive of a short segment IVC occlusion, whose extent was not mentioned. An IVC angiogram showed a complete, blunt obstruction of the IVC, below the IVC-Right Atrial (RA) junction. A simultaneous injection in the IVC-RA junction from the upper limb, revealed complete obstruction of the inferior vena cava by a membrane at 2-3 cm below the IVC right atrial junction.

The patient underwent Percutaneous Translumen Angioplasty (PTA) of the inferior vena caval web. Dilatation was done with 26 mm inoue Balloon and the flow between IVC and RA was established. No significant residual stenosis was seen after the procedure and the collaterals were reduced significantly. The patient was followed up after one month of the procedure, the ascites and engorged veins in the abdomen completely disappeared.

Fig 1: Ultrasonagram

Fig 2: IVC angiogram

Fig 3: Kissing Angiogram

Fig 4: Crossing Membrane

Fig 5: Balloon dilatation

Fig 6: Final angiogram
Discussion
Most of the reports on the membranous obstructions of the IVC, have originated from Japan, India, and south Africa\textsuperscript{9,7}, where this disease has a high incidence. Pathologically, this abnormality has been described as a web or a membrane which obstructs the IVC. Simpson\textsuperscript{7} has described three types of membrane obstructions. In type I, the IVC is obstructed by a thin membrane, at the level of the entrance to the right atrium. In type 2, the segment of the IVC is absent and in type 3, there is complete obstruction of the IVC, which is secondary to the thrombosis. Clinically, these patients present in the third and fourth decades of life, with symptoms of portal hypertension. But our patient presented in the second decade itself. Yang and colleagues\textsuperscript{9} reported good results with percutaneous transluminal balloon membranotomy. Our patient the membranotomy was done by INOUE balloon which is used for balloon mitral commissurotomy. After balloon dilatation if there is no significant residual stenosis then there is no need for stent placement. Srinivas and colleagues\textsuperscript{11} reported that an endovascular management (balloon dilatation ± stenting) for membranous obstruction was safe, with good long-term patency rates. Sinha N and et al\textsuperscript{12}, reported an incidental occurrence of IVC obstruction with Severe Rheumatic Mitral Stenosis for which they did single-staged balloon dilatation of the membranous obstruction. Bansal and et al\textsuperscript{13}., using a Brockenbrough septal puncture needle relieved a membranous obstruction of IVC followed by deployment of a wall stent after series of balloon dilation which yield good results

Conclusion
The rarity in this case is its unusual presentation. Though we could not invasively measure the portal pressure, there was an evidence of portal hypertension as evidenced by ascites. The flow was completely restored. PTA Balloon dilatation with or without stent placement is a viable option for treating membranous obstructions of the IVC, which has shown good long term results.

References


