

The Budd Chiari Syndrome: A 22 year male treated successfully with Mesocaval Shunt

Noureen Malik,¹ Ahsin Manzoor Bhatti,² Syed Mukarram Hussain,³ Junaid Mansoor,⁴ Farrukh Saeed⁵

Abstract

Budd Chiari syndrome (BCS) is a heterogeneous group of disorders characterized by hepatic venous outflow obstruction at any level from the small hepatic veins to the atrio cava junction. BCS could have serious consequences if not treated promptly. Comprehensive angiographic studies, computerised tomography, liver ultrasonography, histology and pathologic analyses are essential for establishing the disease severity, stratifying risk factors, choosing the appropriate therapy and objectively assessing the response. The main objective of therapy is to alleviate portal and IVC hypertension. Here we present a report of a young man with BCS who underwent mesocaval shunt. The eight months follow up showed that patient was free of symptoms and duplex scans confirmed the patency of the shunt.

Keywords: Budd Chiari Syndrome, Hepatic venous outflow obstruction, Mesocaval shunt.

Introduction

Budd-Chiari syndrome (BCS) is the name given to hepatic venous outflow obstruction anywhere from hepatic venules to suprahepatic inferior vena cava (IVC).^{1,2} It usually presents with abdominal pain, distension, hepatomegaly and ascites. BCS needs to be differentiated from common conditions such as Hepatitis and chronic liver disease because if diagnosed early and managed properly patients have fairly good prognosis. Here we present a case of a young man presenting with BCS who was managed medically and then surgically with a mesocaval shunt.

Case Report

A 22 years old male soldier, serving at a height of 19200 feet developed dull abdominal pain, vomiting and mild abdominal distension. He was air-evacuated to Military Hospital Rawalpindi after a week. On physical examination he had mild jaundice, hepatosplenomegaly and moderate ascites. He had no other stigmata of chronic liver disease. Laboratory values revealed normal

blood counts, ALT 121IU/L, serum bilirubin 3mg/dl, Prothrombin time 22/13sec (INR 1.78) and Partial Thromboplastin Time Kaolin 42/34 sec. Ultra sonogram of abdomen showed hepatomegaly (18cm) mild splenomegaly and ascites. Upper GI endoscopy revealed grade 1 oesophageal varices. Duplex scan of abdomen showed thrombosis at hepatic vein confluence with monophasic flow in hepatic veins. Contrast enhanced CT of abdomen revealed characteristic nutmeg/flea-bitten liver with enlarged caudate lobe and nonhomogeneous parenchymal opacification. Thrombophilia, vasculitic screen and hepatitis screening were negative. JAK 2 mutation was also negative. A diagnosis of Budd-Chiari syndrome was therefore made.

He was anticoagulated with enoxaprin 1mg/kg body weight s/c twice a day followed by warfarin 5mg once a day (later on adjusted to keep INR between 2-3). His ascites and splenomegaly regressed but pain in the right hypochondrium persisted and a CT scan done after three months showed some early signs of Cirrhosis. Therefore after a multidisciplinary meeting surgical intervention was planned. This was discussed with the patient and he consented for the operation. Before surgery he had inferior vena caval and right atrial pressure checked to rule out any compression by the caudate lobe. These were within normal limits. The patient underwent a mesocaval shunt with a 8mm reinforced Polytetrafluoroethylene (PTFE) graft. The surgery was straightforward without any untoward incidence and he made an uneventful recovery. Duplex scans done at one and twelve weeks confirmed reverse flow in the portal vein. He was anticoagulated with warfarin 5mg once a day (later on adjusted to keep INR between 2-3). An ultrasound done at eight months revealed some regression in the size of the liver.

Discussion

In 1845, an internist named George Budd described the classic triad of abdominal pain, hepatomegaly and ascites. In 1899, a pathologist Hans Chiari documented the histopathological features of Budd-Chiari syndrome.^{2,3} BCS results from the occlusion of one or more hepatic veins, isolated occlusion of the IVC, the right atrium or the combined occlusion. Around 18% cases have associated portal venous system thrombosis as well.^{1,2} Primary BCS

¹⁻⁴Combined Military Hospital, ⁵Military Hospital, Rawalpindi.

Correspondence: Noureen Malik. Email: noureenmalik66@hotmail.com

originates from within the lumen of the veins or venules and results from thrombus, IVC webs (more common in Asians) or endophlebitis or postoperative complications following liver transplantation.² Secondary BCS results from an extra-luminal lesion such as tumour, abscess or cyst, which can invade the lumen or cause extrinsic compression.⁴

Radiographic analysis should attempt to determine the patency of the hepatic veins, IVC and portal vein because of therapeutic implications. Colour and pulsed Doppler ultrasonography is the first-line investigation for diagnosis with a sensitivity and specificity as high as 85%. CT-scan and MRI may also be helpful. The commonest radiological finding is caudate lobe hypertrophy (72%) followed by hepatomegaly (67%) and splenomegaly (52%). Patients should also undergo testing for hypercoagulable states^{1,5} and bone marrow biopsy (for MPD),^{2,5,6} Infra- and suprahepatic caval pressures, hepatic venography and liver biopsy may aid in management.

The main aim of therapy and interventional management in BCS is to relieve portal and IVC hypertension. Medical management involves anticoagulation and thrombolysis.² Other treatment modalities are percutaneous interventional techniques, surgical shunts and orthotopic liver transplantation (OLT). Local thrombolysis by interventional radiology is preferable to systemic thrombolysis. In acute conditions Catheter-directed thrombolytic Therapy (with tissue plasminogen activator), angioplasty and stent placement (in IVC or hepatic veins) can be helpful. TIPS has become an attractive option in the elective and emergent situations TIPS can be placed through hepatic vein remnant or transcaval. TIPS effectively decompresses the portal system and may serve as a bridge to OLT.^{1,9}

Surgical Shunting procedures are indicated in patients with reversible liver injury.² These surgical shunting procedures make a new connecting channel between hepatic veins and inferior vena cava and affectively lower the portal venous system' blood pressure. For patients without IVC obstruction, the portocaval or mesocaval shunts are reasonable options.¹⁰ For patients with IVC obstruction and/or high infrahepatic to right atrium pressure gradient, mesocaval and portocaval shunts may

not effectively decompress the liver and mesoatrial or mesocaval + cavoatrial shunts are better options.¹⁰ Transplant-free survival rates of 82%, 69% and 62% at 1, 5 and 10 years respectively have been reported.² Our patient did not have any IVC thrombosis and there was no evidence of IVC compression by the caudate lobe. As TIPS was not available locally and patient was young so we decided to go for a mesocaval shunt.

Conclusion

BCS is a rare condition which presents with fairly common symptoms. A high index of suspicion with corresponding investigations can diagnose the condition. Complete work-up for thrombophilia including screening for MPD should be carried out which should not stop after identification of a single cause, because almost half of patients have more than 1 underlying disorder. BCS patients have excellent 1- and 2-year survival rates when adequately managed.

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