BUDD CHIARI SYNDROME WITH HEPATO HYDROTHORAX

Col SC BHATTACHARYA *, Lt Col R ANAND +,
Lt Col AP RADHAKRISHNAN +, Lt Col GSN MURTHY #

MJAFI 1999; 55 : 357-358

KEY WORDS: Budd-Chiari syndrome; Percutaneous transhepatic venography; Refractory hepato-hydrothorax; Veno-angiographic balloon dilatation and stenting.

Introduction

Budd-Chiari Syndrome (BCS) comprises of hepatomegaly, abdominal pain, ascites and hepatic histology showing zone 3 sinusoidal distension and pooling [1]. The syndrome is a consequence of hepatic venous obstruction and a variety of conditions have been implicated in its causation ranging from membranous webs in the supra hepatic segment of the inferior vena cava (IVC), to constrictive pericarditis or Right heart failure. Various thrombotic conditions such as myeloproliferative disorders, particularly Polycythemia rubra vera, Systemic lupus erythematosus with circulating lupus anti-coagulant, antithrombin III deficiency and oral contraceptive use can result in hepatic vein thrombosis. Thrombosis in IVC secondary to adrenal or renal malignancy as well as obstruction from direct invasion by hepatocellular carcinoma and angiosarcoma have been reported. However, in 25% cases, the cause remains obscure. We describe here, an unusual presentation of a case of BCS who presented with refractory pleural effusion without ascites or abdominal pain and who left a trail of presumptive diagnoses before being successfully managed by interventional radiology at an International Medical Centre, USA.

Case Report

A 52-years-old Ordnance Factory employee with no significant past medical history presented in Jan 98 with increasing dyspnoea, along with weight and appetite loss. He denied fever, chills, night sweats, cough, abdominal pain, haematemesis or melena. He was a non-alcoholic. Clinical examination revealed an anicteric patient with Rt sided pleural effusion and 2 cm non tender hepatomegaly. There was no ascites and spleen was not palpable. Blood counts were normal and serum transaminases were marginally elevated. Serum bilirubin was 1.1 mg%. Chest skiagram showed a massive Rt pleural effusion. Analysis of pleural fluid revealed protein content of 1.6gm%, RBC 150/cumm, WBC 200/cumm with 91% lymphocytes and atypical mesothelial proliferation which raised suspicions of malignancy. CT chest showed compressive collapse of Rt lung from pleural effusion but without evidence of any parenchymal abnormality. USG abdomen showed an enlarged liver with

heterogenous texture and without any focal mass. Spleen measured 12 cm and there was no ascites. The patient underwent thoracentesis on three occasions within 48 hours of admission as re-accumulation of fluid was found to occur almost immediately resulting in unacceptable dyspnoea. On the surmise that we may be dealing with a case of pleural mesothelioma, or a hepato-hydrothorax secondary to chronic liver disease he was transferred to Cancer institute at Mumbai. Evaluation of serial pleural fluid cytology and pleural biopsy conclusively excluded malignancy. Hepatic scintiscan (99TC) showed poor parenchymal uptake with partial cold areas and border line splenomegaly. The presence of Gradel-II oesophageal varices on UGI endoscopy along with marginal transaminase elevations was again suggestive of chronic liver disease. FNAC liver done to exclude infiltrative disease revealed lymphocytes and scattered polymorphs. A presumptive diagnosis of Hepato-hydrothorax was considered alongwith the possibility of tuberculosis for which 4 drug ATT started was continued. He was tapped dry on several further occasions with recurrence of effusion each time. A second opinion was hence sought and the patient was flown to USA for evaluation at the Medical Centre, University of San Francisco, California. After initial workup at the Pulmonology department which also confirmed the absence of malignancy, he underwent 3 cycles of pleurodesis with talc which proved refractory due to rapid re-accumulation of effusion. Hence a Hepatology consultation was sought. Abdominal ultrasound with Doppler imaging revealed interesting findings which were consistent with BCS. The middle hepatic vein was segmentally occluded with reversal of flow direction through communicating channels to the right hepatic vein.

Fig. : Follow up ultrasound scan showing patent stent in the middle hepatic vein adjacent to the inferior vena cava

* Senior Adviser (Medicine), + Classified Specialist (Medicine), # Classified Specialist (Radiology) Military Hospital, Jabalpur.

The left hepatic vein was not visualised and the left lobe was diminutive as against an enlarged caudate lobe. Both right and left portal veins were of normal calibre but, appeared to have slow flow. There was no ascites. Using the transfemoral route, digital subtraction inferior vena-cavography following selective catheterisation of right and middle hepatic veins to delineate the obstruction from below, was carried out. The middle hepatic vein could not be negotiated by the guide wire, which was then done by percutaneous transhepatic venography (PTHV). This successfully delineated the presence of high grade focal stenosis at its junction with the IVC. The right hepatic vein also showed focal segmental occlusion distally.

The left hepatic vein was rudimentary, ending in a blind stump. Next, an 8 mm x40 mm angioplasty balloon catheter was introduced trans-jugularly to dilate the stenosis of the middle hepatic vein at its junction with the IVC. The existing pressure gradient of 27 mm Hg between the middle hepatic vein (32 mm Hg) and right atrium (5 mm Hg) was reduced initially to 16 mm Hg. Following deployment of an 8 mm x 40 mm stent in the distal middle hepatic vein the pressure gradient was further reduced to a final of 9 mm Hg. Post procedure, the patient had dramatic relief from dyspnoea with no recurrence of pleural effusion or development of ascites. The patient was discharged after 48 hours with removal of chest tube and discontinuation of ATT. He was advised aspirin in anti-platelet doses and Doppler ultrasound evaluation every 3 months for one year and half yearly thereafter.

Discussion

The liver is a discrete organ whose functions depend upon the integrity of cellular, biliary and vascular processes. BCS is a manifestation of hepatic venous flow obstruction leading to tender hepatomegaly, ascites and non specific elevation of hepatic enzymes [2]. Our patient was remarkable by the absence of ascites and presence of a rapidly re-accumulating right sided hydrothorax which proved refractory to pleurodesis. Work up had excluded pleural based infection, malignancy and hypercoagulable states. The phenomenon of right hepato-hydrothorax in the absence of ascites is seen on rare occasions on account of the negative intrapleural pressure during inspiration sucking in peritoneal fluid through diaphragmatic defects into the pleural cavity [3]. A literature search in the INTERNET could not reveal any case where hepatic hydrothorax was reported in hepatic vein stenosis (Budd Chiari syndrome with hepato hydrothorax).

In view of scintiscan findings of non homogenous hepatomegaly with borderline splenomegaly, Gr I-II varices on UGI endoscopy and marginal transaminase elevations, the possibility of hepato-hydrothorax secondary to chronic hepato-cellular disease was first considered. That we were dealing with a primary vascular problem was conclusively shown by Doppler study as evidenced by reversal of hepatic venous blood flow. Transfemoral digital subtraction inferior vena cavography failed to delineate the exact site and

extent of venous obstruction. This was made possible only by PTHV which also showed a rudimentary left hepatic vein with an atrophic left lobe indicative of a more chronic aetiology. Angiographic studies were in favour of a possible web which is seen more frequently amongst patients of BCS in Japan, South Africa and India [4]. PHVT is now being recognised as a standard angiographic procedure once preliminary transfemoral inferior vena cavography and selective hepatic venography fail to delineate the extent of venous obstruction. Wilson et al [5] have demonstrated the usefulness of this procedure in planning out treatment strategies on six patients of BCS. One patient with intraluminal thrombus in right and middle hepatic veins was treated with fibrinolytic infusion and balloon thrombectomy. Two patients having central obstruction of right hepatic vein were treated with venoplasty and stent placement. Three patients showed a "spider web" appearance with diffuse obliteration of the normal intra hepatic venous architecture and for whom a transjugular intrahepatic portosystemic shunt (TIPS) was placed.

Our patient responded well to dilatation and stenting resulting in a rapid reduction of the pressure gradient between the hepatic veins and IVC. Fortunately stenting could be carried out successfully as there was a sufficient vessel remnant of the middle hepatic vein. Should this not have been so, a TIPS procedure could have been necessary with all its attendant complications. The patient has been enjoying good health till date. Periodic Doppler ultrasound evaluations carried out in this Hospital have shown stent patency. Follow up liver function tests are consistently within normal limits.

REFERENCES

1. Sherlock S, Dooley J. Disease of the liver and Biliary System, 10th ed. Oxford : Blackwell Science Ltd.
2. Mitchell MC, Boitnott JK, Kaughan S, et al. Budd-Chiari Syndrome: Etiology, diagnosis and management. *Medicine* 61;199:1982.
3. Giacobba A, Faccio RD, Tonti P, et al. Hydrothorax complicating cirrhosis in the absence of ascites. *J Clin Gastroenterology* 1993;16:271.
4. Myung HH, Hahn PY, Gadallah SF, Crockett J. Hepatic hydrothorax: Possible etiology of recurring pleural effusion. *American Family Physician* August 1997;33(8).
5. Valla D, Benhamou JP. Obstruction of the hepatic veins or supra-hepatic vena cava. *Dig Dis* 1996;14:99.
6. Wilson MW, Ring EJ, LaBerge JM, Kerlan RK, Gordon RL. Percutaneous transhepatic venography in the delineation and treatment of Budd-Chiari Syndrome. *J of Vascular and Int. Radiology*. 1996 Jan-Feb; 7(1):133-8.