International Journal of Allied Medical Sciences and Clinical Research (IJAMSCR)

ISSN:2347-6567

IJAMSCR |Volume 5 | Issue 3 | July - Sep - 2017
www.ijamscr.com

Research article *Medical research*

An interesting case of ascites – budd chiari syndrome

Karthick Ramalingam[1], Mariraj I[2], Jagadeesan M[2*], Mahendra Kumar K[2], Prasanna karthik S[3], Kannan R[4], Magesh kumar S[4]

[1]*Assistant Professor, Department of General Medicine, Sree Balaji Medical College Hospital, Bharat University, Chennai, Tamilnadu, India*

[2]*Assistant Professor, Department of General Medicine, Saveetha Medical College Hospital, Saveetha University, Chennai, Tamilnadu, India*

[3]*Associate Professor, Department of General Medicine, Saveetha Medical College Hospital, Saveetha University, Chennai, Tamilnadu, India*

[4]*Professor, Department of General Medicine, Saveetha Medical College Hospital, Chennai, Saveetha University, Chennai, Tamilnadu, India*

***Corresponding Author: Dr. Jagadeesan M**

Email: drjagadeesan@gmail.com

ABSTRACT

Budd-chiari syndrome is defined as the obstruction of hepatic veins or terminal inferior vena cava (IVC) [1-3].It is a rare disease and can be life-threatening. BCS can be classified as primary when BCS arises from a venous anomaly or as secondary, when an initial lesion is outside the veins. The incidence may be higher in Asian than western population with a slight female preponderance. Hereditary thrombophilia is a condition in which many inherited conditions leading to an increased tendency to develop venous thromboembolism (VTE). Those conditions are very rare. As such BCS is not common in these conditions [3]. In this case report we present a case of Budd- Chiari syndrome with more than one such risk factors.

Keywords- Budd chiari, BCS, Protein C, Hepatic vein

CASE PRESENTATION

A 20 year old female presented with abdominal distension for 10 days. It is insidious in onset and progressive in nature. Patient complained of high colored urine. Patient had right upper abdominal pain which was dull aching in character. There was bilateral pitting pedal edema and decreased urine output. There was no history of chest pain, palpitation, sweating, or syncope. There was no history of fever, cough with expectoration, vomiting, hematemesis or malena. Patient had delivered a healthy female baby five months ago. There was no history of oral contraceptive pill intake or any other drug intake. There was no significant past medical history including liver stigmata. Patient did not use illicit drugs and there was no identifiable high risk sexual behavior. On clinical examination, patient was pale and icteric. Bilateral pitting pedal edema was present. Other stigmata of chronic liver disease were not present. Abdominal examination revealed ascites and tender

hepatomegaly extending 3 cm below the right costal margin. Spleen was just palpable. Hepatojugular reflex was absent. There were no dilated veins seen over the abdominal wall. Laboratory tests revealed normal blood counts except mild anemia. Renal function and echocardiogram were normal. Serum total bilirubin was 3.1 mgs%, ALT was 121 IU/L, and AST was 128 IU/L. ALP was normal. USG abdomen confirmed the clinical findings and portal venous doppler study revealed absent flow in hepatic veins and IVC. Portal vein flow was normal. Her bone marrow examination was found to be normal. CECT abdomen confirmed the same and diagnosis of Budd-Chiari syndrome was made. A battery of investigations was done to identify the cause of BCS. Levels of Protein C, Protein S and Antithrombin III were significantly low. Protein C level was 24% (normal-70% to 130%), Protein S level was 49% (normal-72% to 116%) and Antithrombin level was 34.6% (normal-80 to 120%). Other investigations including APLA, factor V laiden mutation, VDRL, homocysteine levels were done and found to be negative. Patient was started on warfarin after combining it with heparin. INR level was maintained around 2.5. Patient showed dramatic clinical improvement with improvement in flow in both IVC and hepatic veins.

DISCUSSION

In most instances the cause of BCS is idiopathic; however, particular causes to be noted are IVC webs, hypercoagulable states, metastatic neoplasm, infection or trauma [table1].In a study by Darwish et al , it was found that one risk factor was seen in 84% of patients and two or more risk factors in 48% [4]. Myeloproliferative disorders are the most common cause of primary type, of which Polycythemia Rubra Vera (PRV) is the most common. Secondary BCS can be caused by several mechanisms including (i) invasion by malignancy or infectious process (ii) compression by focal nodular hyperplasia or cysts (iii) compression and inflammation by liver abscesses or polycystic liver disease and (iv) blunt trauma [5]. Part of liver drained by at least 2 hepatic veins should be obstructed before the clinical features develop [5]. The cascade of events following the development of obstruction resulting in pressure and blood flow changes leading to development centrilobular necrosis in liver as well as formation of ascites. Liver region with intact blood flow undergo hypertrophy as in segment l (speigel lobe).

Fibrosis resulting from necrosis typically result in venocentric cirrhosis. This can result in fulminant liver failure as well as portal hypertension.

Table 1 – Etiology of Budd Chiari syndrome

Hypercoagulable States
Antiphospholipid syndrome
Antithrombin deficiency
Factor V Leiden mutation
Methylenetetrahydrofolate reductase TT677 polymorphism
Myeloproliferative neoplasm
Oral contraceptives
Paroxysmal nocturnal hemoglobinuria
Postpartum thrombocytopenic purpura
Pregnancy
Protein C deficiency
Protein S deficiency
Prothrombin gene mutation G20210A
Sickle cell disease
Infections
Aspergillosis
Filariasis
Hydatid cysts (*Echinococcus granulosus* or *E. multilocularis*)
Liver abscess (amebic or pyogenic)

Pelvic cellulitis
Schistosomiasis
Syphilis
TB
Malignancies
Adrenal carcinoma
Hepatocellular carcinoma
Leiomyosarcoma
Leukemia
Lung cancer
Myxoma
Renal carcinoma
Rhabdomyosarcoma
Miscellaneous
Behçet's disease
Celiac disease
Dacarbazine therapy
IBD
Laparoscopic cholecystectomy
Membranous obstruction of the vena cava
Polycystic liver disease
Sarcoidosis
Trauma to abdomen or thorax

The clinical features can be chronic, sub acute (more common types), or acute (less common types). Usual symptoms are abdominal pain, ascites and fever. Stigmata of chronic liver disease including portal hypertension, GI bleeding, hepatorenal syndrome, hepatic encephalopathy and bacterial infections can occur. Liver function tests may be altered to varying degrees [4]. Hypersplenism arising from portal hypertension can normalize the cell counts in myelo proliferative disorders [6].

In our patient there was no evidence of chronic liver disease portal hypertension. Because coagulation inhibitors are decreased nonspecifically by liver disease, a primary deficiency in protein C, protein S, or antithrombin is difficult to establish [3]. As our patient's liver was normal, primary deficiency of these factors was made. Patient was started on warfarin and overlapping with heparin was done. After 4 weeks of therapy, there was partial recanalisation of IVC and hepatic vein.Anticoagulation and medical therapy remains the first line of management in BCS. Intervention and surgical treatment are reserved for patients with a fulminant course or for those not responding to medical therapy [7].

CONCLUSION

We report this case because of the rarity of the condition and the unusual cause underlying it. Patients with isolated protein C may remain asymptomatic or present with thromboembolic incidents in the event of precipitating factors. Early diagnosis, appropriate management, long-term prophylaxis and proper counseling can prevent recurrent thromboembolic events and reduce the long-term morbidity. This case signifies the importance of looking for multiple risk factors for BCS, as the patient can have more than one cause.

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How to cite this article: Karthick Ramalingam, Mariraj I, Jagadeesan M, Mahendra Kumar K, Prasanna karthik S, Kannan R, Magesh kumar S. An interesting case of ascites – budd chiari syndrome. Int J of Allied Med Sci and Clin Res 2017; 5(3): 767-770.
Source of Support: Nil. **Conflict of Interest:** None declared.