

Budd Chiari Syndrome Presenting As Pelvic Congestion Syndrome : A Rare Case Report

Dr. Shrivastava Priyanka¹, Dr. Mohan Parveen², Dr. Santpur Unmesh³

¹Rims, Ranchi

²Mmimsr, Mullana, Ambala

³Mmimsr, Mullana Ambala

Abstract

We present a young female patient who had pelvic congestion syndrome caused by pelvic varicosities associated with Budd-Chairi syndrome, but without other symptoms or signs related to Budd-Chairi syndrome. Medical therapy was effective in improving her symptoms.

Keywords: Pelvic congestion, Budd-Chiari syndrome

I. Introduction

Following the initial report of Budd in 1845 and Chiari in 1899 on the occurrence of occlusion of hepatic veins, a number of reports have appeared about different aetiological factors in the genesis of Budd-Chairi syndrome (BCS) and its various modes of presentation. It may present with the classical features of ascites, jaundice, hepatomegaly and/or variceal bleeding or clinical signs caused by venous blood retuning via alternate routes to the heart may be the sole presentation¹. However, we recently came across a young female patient who had pelvic congestion syndrome due to chronic BCS which is a rare occurrence.

II. Case Report

We report a case of a 30 year old housewife who presented in the Gynae OPD with the chief complaints of visible veins over the abdomen and chest for over 8 years, chronic pelvic pain, secondary dysmenorrhea and menorrhagia for 2 years. She had two children. There was no history of pain in the abdomen, jaundice, or febrile episodes. Physical examination revealed a well built woman of average nutrition. Prominent, dilated, tortuous superficial veins were present over the abdomen and the chest with blood flow from below upwards. The liver which was enlarged by 3 cm below the costal margin, was firm and non-tender. Pelvic examination revealed a normal-sized uterus with no adnexal lesion. Results of investigations did not show any abnormality in liver function tests. Ultrasound examination of the abdomen revealed attenuated left and right hepatic veins with visible multiple intrahepatic collaterals (figure 1), suggestive of BCS while ultrasound of the pelvis was normal. On Color doppler there was reversal of flow in hepatic veins (figure 1). Contrast enhanced computer tomography (CECT) of the abdomen and pelvis showed blockage of the suprahepatic part of IVC with dilation of azygous and hemizygous veins and the pelvic veins (figure 2). Therefore, the diagnosis of pelvic congestion syndrome was made, caused by inferior vena cava reflux associated with BCS. She was started on Medroxy Progesterone Acetate (MPA) 10 mg twice daily for twenty one days. Patient showed symptomatic improvement on follow up for 3 months.

III. Discussion

BCS is a heterogeneous group of disorders characterized by hepatic venous outflow obstruction at the level of hepatic venules, large hepatic veins, inferior vena cava (IVC) or the right atrium². Pelvic congestion syndrome (PCS) appears as the very variable combination of chronic pelvic pain, dyspareunia, dysmenorrhea, menorrhagia, urinary symptoms (e.g. dysuria, pollakiuria, bladder urgency) and rectal symptoms (e.g. constipation)³.

Depending upon the site of obstruction, BCS is classified as under²:

- Small hepatic veins
- Large hepatic veins
- Inferior Vena Cava
- Combined obstruction-both hepatic veins and IVC

Our patient had combined obstruction, which is the commonest disease pattern in cases of BCS reported from India⁴. Patients with the acute form of the disease usually presents with ascites, hepatomegaly, jaundice and / or esophageal / gastric variceal bleeding. Though BCS usually presents with symptomatic

disease, less commonly asymptomatic disease is detected on imaging done for some other indication⁵. Chronic venous congestion of reproductive organs may be another presentation⁶ as in our case where the patient had long standing chronic IVC obstruction which lead to pelvic varicosities because pelvic structures are drained by both internal iliac veins and genital veins.

Varicose veins can develop by two pathophysiological mechanisms:

1. Reflux due to valvular incompetence
2. Obstructive diseases e.g. Nutcracker Syndrome, Post-thrombotic disease involving the common iliac vein and/or the IVC, BCS³.

Some authors have suggested partial suppression of ovarian function with MPA may relieve symptoms of chronic congestion³ which was done in our case as patient could not immediately afford surgical treatment. Embolisation of the pelvic veins or surgical stenting of the thrombosed veins provide better long term results. The main complications of Budd–Chiari syndrome have been described in various patient series and case reports. However, the rarity of the syndrome hinders extensive studies on prognostic factor.

IV. Conclusion

BCS may manifest in various forms so its diagnosis should be considered even if obvious signs are absent.

References

- [1]. Madangopalan N, Soloman V, Jayanthi V, et al. Clinical spectrum and surgical relief of Chronic obstruction of hepatic and suprahepatic segment of inferior venacava of chronic Budd Chiari Syndrome. J Gastroenterol Hepatol 1986; 1: 359 -69
- [2]. Ludwig J, Hashimoto E, McGill DB, van Heerden JA. Classification of hepatic venous outflow obstruction: ambiguous terminology of the Budd-Chiari syndrome. Mayo Clin Proc 1990;65:51
- [3]. Hartung O. Pelvic varicose veins:Embolisation by vascular surgeon.Controversies and updates in vascular surgery 2006.Available from URL (www.cacvs.org/.../EN/.../HARTUNG_BARTHELEMY_BOUFI_ALMI.pdf)
- [4]. Eapen C E, Thomas Mammen, Vinus Moses, et al. Changing profile in Budd Chiari syndrome in India. IJG 2007 ;26 :77-81
- [5]. Hadengue A, Poliquin M, Vilgrain V, et al. The changing scene of hepatic vein thrombosis: recognition of asymptomatic cases. Gastroenterology 1994;106:1042-7.
- [6]. Victor S, Jayanthi V, Madanagopalan N.Coarction of the inferior vena cava. Trop Gastroenterol 1987;8:127-42

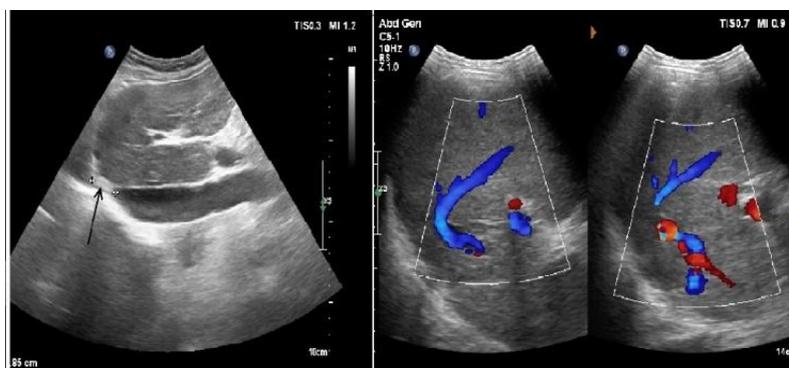


Fig 1 : Usg Image (Left) Is Showing Obstruction Of The Suprahepatic Ivc (Arrow). Color Doppler (Right) Is Showing Intrahepatic Venous Collaterals And Reversal Of Flow In Hepatic Veins .



Fig 2 : Coronal Cect Image (Left) Is Showing Dilated Pelvic Veins (Arrow) Suggestive Of Pelvic Congestion. Axial Cect(Right) Is Showing Dilated Hemiazygous Vein (Arrow), Dilated Abdominal Wall Collaterals(White Dotted Arrow), And No Flow In Ivc (Black Dotted Arrow)