BUDD CHIARI SYNDROME - A CASE REPORT

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ABSTRACT

BUDD CHIARI SYNDROME results from obstruction to the venous outflow of liver from the hepatic veins till the inferior vena cava. Clinical presentation of Budd Chiari Syndrome is variable. Most of the patients present with abdominal pain, hepatomegaly and ascites. We report case of 44yrs female admitted in hospital for the evaluation of Budd Chiari Syndrome. The importance of correct diagnosis by the physician and subsequent management is reviewed.

KEY WORDS: Budd Chiari Syndrome, myeloproliferative disorders, thrombotic, non-thrombotic

I. INTRODUCTION

Budd-Chiari syndrome (BCS) was actually described as an obstruction of hepatic venous outflow or hepatic portion of the inferior vena cava (IVC) [1]. The symptoms resulting from this type of occlusion of the hepatic outflow, “classical BCS”, were first described by Budd [2, 3] in 1845 and later by Hans Chiari in 1899. It has been described to occur in 1 in 100,000 of the population [4, 5]. This syndrome usually occurs when a clot narrows or blocks the hepatic veins, which carry blood out of the liver. Because blood flow out of the liver is impeded, blood backs up in the liver, causing it to enlarge and the spleen may also enlarge. This causes blood pressure in the portal vein (which carries blood to the liver from the intestines) to increase. This increased pressure, called portal hypertension, can result in dilated, twisted (varicose) veins in the esophagus (esophageal varices). Portal hypertension, plus the damaged liver, leads to fluid accumulating in the abdomen (called ascites). The kidneys contribute to ascites by causing salt and water to be retained. The clot may extend and also block the inferior vena cava (the large vein that carries blood from the lower parts of the body, including the liver, to the heart). Varicose veins in the abdomen near the skin’s surface may develop and become visible. Finally, severe scarring of the liver (cirrhosis) occurs [6]. Symptoms may vary from a completely asymptomatic condition to fulminant liver failure [7]. The diagnosis of Budd-Chiari syndrome should be clinically diagnosed in patients who present with any one of the following findings. They are fulminant liver failure with abrupt onset of ascites and hepatomegaly, massive ascites with relatively preserved liver function, unexplained chronic liver disease, or liver disease and an associated thrombogenic disorder [8]. Budd-Chiari syndrome may be classified into three types depending on the type of existing venous occlusion. Type I is limited to the inferior vena cava. In type II BCS, lesions are within the hepatic veins. If lesions are short-segment occlusions (< 4 cm), type II a Budd-Chiari syndrome is the diagnosis. Budd-Chiari syndrome type III is the mixed type with involvement of the IVC and hepatic veins. Imaging studies play an important role in confirming the diagnosis.
of Budd-Chiari syndrome by showing the venous abnormalities. Probably the most useful imaging methods include conventional and Doppler ultrasound, CT, MRI, and catheter venography [9]. The management of Budd-Chiari syndrome can be divided into three main categories: medical, surgical, and endovascular [10-13].

II. CASE REPORT

A female patient of age 44 yrs was admitted in hospital with the complaints of severe abdomen pain and loss of appetite generalized weakness since 15 days. Past history was Known Case of post MHV stenting 8yrs back. Her laboratory investigations include Hemoglobin:11gm%; WBC: 1700 thousands/cumm; platelets :1.5 lakhs; Na:137 mmol/lit Ca:108mg/dl; K : 3.6 mmol/lit; Total Bilirubin :0.7 mg/dl; Direct Bilirubin :0.6 mg/dl; Ultra Sonograph of abdomen: Intra hepatic IVC narrowing. Hemoglobin :11gm%; WBC: 1700 thousands/cumm; platelets :1.5 lakhs; Na:137 mmol/lit Ca:108mg/dl; K : 3.6 mmol/lit; Total Bilirubin :0.7 mg/dl; Direct Bilirubin :0.6 mg/dl; Ultra Sonograph of abdomen: Intra hepatic IVC narrowing. In June 2006 the patient was operated and had Transp 2006; 12(suppl 2):S23

CONCLUSION

Budd-Chiari syndrome is a rare case in females this syndrome can be fast and lead to death in couple of months and may be considered sudden onset of abdominal pain and hepatomegaly. After diagnosis antithrombotic medications must be given immediately.

V. REFERENCES

[6] Full review/revision October 2018 by Nicholas T. Orfanidis, MD

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