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Case Report

CASE REPORT ON BUDD CHAIRI SYNDROME

Rahul K*., Soundharya Lakshmi R and Sai Reddy S

Department of PharmD, Smt. Sarojini Ramulamma College of Pharmacy, Mahabubnagar
Department of Pharmacy Practice, Smt. Sarojini Ramulamma College of Pharmacy, Telangana State, INDIA

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ABSTRACT

Budd-Chairi Syndrome is a rare condition characterized by obstruction of venous outflow through hepatic veins which drains the liver. This is a case report of 35years female patient who developed Budd-Chairi Syndrome. Patient presented to the hospital with less haemoglobin levels (anemia), pain and swelling of the abdomen. Past history of spontaneous abortion of 1st pregnancy at 2nd month and has undergone regular ANC checkup's. Menstrual history includes menarche at 12yrs, MH-5/30days, regular, no clots. For diagnosis, lab investigations were performed and from the obtained results it was confirmed as Budd-Chairi Syndrome. The treatment given was accordingly with anticoagulants and antibiotics and the patient was relieved from symptoms. The main intension of our work is to bring awareness among health-care professionals about the disease and its complications.

INTRODUCTION

Budd-Chairi syndrome is a rare condition having prevalence of 1 in a million individuals[1]. BCS is characterized by hepatic venous outflow obstruction extending at any level from small hepatic veins to the atriocaval junction [2]. Etiological factors include hematological disorders, pregnancy, contraceptive pill, and trauma. Common thrombophilic factors associated with BCS include antithrombin deficiency, protein C deficiency, protein S deficiency, heterozygous factor- Factor V Leiden, prothrombin G20210A mutation [3] The classical triad of abdominal pain, ascites, and hepatomegaly observed in majority of patients and are asymptomatic. The clinical variants include: Acute and Sub acute, Chronic, Fulminant [4]. The disorder can be suspected in any patient who develops massive ascites, the diagnosis can be confirmed accurately by hepatic venography [5]. The pathophysiological changes involve obstruction generally caused due to thrombus, or may result for extrinsic constriction such as tumors, abscess, cysts or formation of membranous webs within the inferior vena cava (IVC)[2]. The mutation in the prothrombin G20210A gene, causes 25% increase in plasma prothrombin levels causing increased risk of thrombotic events [6]. Due to the obstruction of the portal vein there is an increased sinusoidal pressures which eventually leads to: i) increased filtration of fluids causing ascites, ii) collateral venous flows through alternate veins causing gastric varices. If left untreated leads to hepatic necrosis and eventually to liver failure. In pregnancy Factor II gene mutation was responsible for poor outcome. A retrospective study reports of sixteen women in 24 pregnancies with BCS having treated with anticoagulants showed better outcome[7]. Early Diagnosis and proper treatment would be beneficial for the good results. In this case pregnancy would be the cause of the Budd-Chairi Syndrome. Treatment strategies of BCS generally follow least invasive to most invasive which include anticoagulant therapy, anti-fibrinolytics, diuretics and surgical treatment such as TIPS, liver transplantation [8].

CASE REPORT

A female patient aged 35years was admitted to hospital with chief complaints of G_2A_1 with less hemoglobin count (moderate anemia), abdominal pain and swelling. She had a previous history of spontaneous abortion at 2nd month. Her Menstrual history includes Menarche at 12yrs, MH- 5/30days, regular, no clots, no dysmenorrheal Condition. She had a past history of germ cell tumor (ovary) at 14yrs of age taken chemotherapy. Treatment was given accordingly to relieve her present complaints and ANC's were done at regular intervals and was asked her to visit the hospital at next conception and she did.

Corresponding author:* **Rahul K
Department of PharmD, Smt. Sarojini Ramulamma College of Pharmacy, Mahabubnagar

Laboratory Investigations

- Physical examination: represents normal blood pressure- 120/80mmHg (120/80mmHg), swelling of abdomen.
- Complete Blood Picture revealed decreased levels of Hemoglobin- 8.0gm/dl (11.0-16.0gm/dl), RBC count- 3.5mill/cu mm (4.2-6.8mill/cu mm).
- Liver Function Test showed impaired levels of Total Bilirubin- 1.9mg/dl (0.22-1.0mg/dl), Direct Bilirubin- 0.8mg/dl (0.02mg/dl), SGPT- 155U/L (0-48U/L), Albumin- 2.3gm/dl (5.5-8.0gm/dl), Globulin- 3.1gm/dl (3.5-5.0gm/dl), Alkaline phosphate- 180U/L (20-125U/L).
- Increased Prothrombin time- 2.1(INR-0.8-1.1), APTT- 32.7sec (22-32sec).
- Renal Function Test shows normal levels of Urea- 39mg/dl (15-40mg/dl), serum creatinine- 1.5mg/dl (0.6-1.3mg/dl).

Differential Diagnosis

USG Abdomen reveals thrombosis, massive ascites. Decreased Hemoglobin levels represents moderate anemia. Correlating her previous history and laboratory test results the physician diagnosed as Moderate Anemia with BUDD-CHAIRI SYNDROME.

Treatment

Patient was given following medications on admission Inj. Inhep 2ml/OD, Tab. Dytor 10mg/OD, Inj. Taxim-1gm/ BD, Tab. Pantop- 20mg/BD, Inj. Human albumin-20gm/OD, Tab. Neurokind- 1500mcg/OD, protein powder TID, Electrol powder. The same treatment was performed for seven days, and then the patient was relieved from symptoms of BCS. Regular monitoring of the patient is required to avoid further complications.

RESULTS AND DISCUSSION

Budd-Chairi Syndrome is characterized by decreased venous outflow due to obstruction of hepatic vein which drains the liver. The patient came to hospital for ANC.She has some complaints at her last visit to hospital. She already lost 1 pregnancy due to BCS. The treatment was given accordingly to reduce the symptoms of BCS.

Table 1 Investigations to confirm Budd-Chairi Syndrome

Sl. No.	Investigations	In present case	Normal range
1	Physical examinations	Swelling in the abdomen	-
2	Complete Blood Picture		
	Hemoglobin	8.0gm/dl	11.0-16.0gm/dl
3	Liver Function Test		
	Total Bilirubin	1.9mg/dl	0.22-1.0mg/dl
	Direct Bilirubin	0.8mg/dl	0.02mg/dl
	SGPT	155U/L	0-48U/L
	Albumin	2.3gm/dl	5.5-8.0gm/dl
	Globulin	3.1gm/dl	3.5-5.0gm/dl
	Alkaline Phosphate	180U/L	20-125U/L
4	Prothrombin Time	2.1	INR-0.8-1.1
	APTT	32.7sec	22-32sec
5	Ultrasonography	Thrombus, Massive ascites	–

Heparin was given to the patient as part of treatment to lyse the clot so that the blood flow attains normal their by decrease the sinusoidal pressure in that patient.

Due to administration of above therapy, the delivery was successful and safe. Mother is monitored regularly as she may still present underlying prothrombotic condition. Patient can be advised for surgical treatment like TIPS which has got a good clinical outcome [9].

CONCLUSION

The reliable characteristic features may also be present after the initial therapy of anticoagulants. The treatment given to the patient was safe and effective but delivery was done in 7th month of the pregnancy to avoid further complications. . The main intension of our work is to bring awareness among health-care professionals about the disease and its complications.

Abbreviations Used

ANC- Ante Natal Care
MH- Menstrual history
$_{G2A1}$-Second gravid [Pregnancy]with previous history of abortion[$_{A1}$]
IVC- Inferior vena cava
APTT-Activated Partial Thromboplastin Time
USG- Ultrasonography
TIPS- Transjugular Intrahepatic Portosystemic Shunt

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