Case Report on "Budd Chiari Syndrome"

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Abstract

Background: Hepatic Vein Thrombosisis also known as Budd Chiari Syndrome is a sparse disorder marked by obstruction and narrowing of the hepatic veins i.e., veins of the liver. 70% casers of Budd Chiari Syndrome are unknown. Approximately 10% have polycythemia vera which is a type of blood cancer. Clinical manifestations can be developed due to the blockage of the major veins which carries blood from the liver to the heart. Features may include hepatomegaly (an abnormally enlarged liver), pain in the upper right part of the abdomen, yellowish color of the skin and sclera (whites of the eyes) jaundice i.e., due to jaundice, ascites i.e., accumulation of fluid in the peritoneal cavity between the two layers of membrane that lines the stomach. Portal hypertension (due to increase in pressure of the portal veins) etc. Diagnosis of hepatic vein thrombosis is made based upon a detailed diagnostic findings, a thorough history of patient, and a variety of specialized tests. Management of hepatic vein thrombosis includes corticosteroid drug. anticoagulants, angioplasty, etc. **Patient history:** The patient is a female, 46 years old who was admitted in A.V.B.R.H. on 29/11/19 with chief complaints of tenderness in abdomen on palpation, fever, rigor and pain. **Past history:** Patient is a known case of hypertension and cerebrovascular accident. **Clinical findings:** The patient has undergone venography, Complete blood count, etc. **Pharmacology:** The patient was treated with medications such as, tab. Augmentin 625 mg, tab. Pantop 40 mg, tab. Ultracet 50 mg, tab. Mucomix 625 mg. **Nursing management:** Administered IV fluids as advised by physician. Administered medications as prescribed by physician. Monitored vital signs. **Conclusion:** The 46 years old female was admitted to AVBRH in Cathlab ICU with chief complaints of pain in abdomen, fever, rigor etc. after undergoing investigations she was diagnosed as Budd Chiari Syndrome.

Keywords: *Budd Chiari Syndrome, Hepatomegaly, Ascites, Cirrhosis of Liver.*

Introduction

Hepatic venous thrombosis or Budd Chiari Syndrome (BCS) is a very infrequent condition, which affects one in a million. BCS or hepatic venous thrombosis is a condition which occurs due to the occlusion of the hepatic veins which drains the liver. It is formally introduced with the classical triad of ascites (fluid collection in the peritoneal cavity), pain in abdomen and enlargement of the liver.(1) The causes of Budd Chiari syndrome is classified as: 1. Primary cause of Budd Chiari syndrome (which contributes to 75 %) includes, Thrombus of the veins in liver. It may be associated with following disorders: Polycythemia vera, Pregnancy, Postpartum state, Use of oral contraceptives, Paroxysymal nocturnal hemoglobinuria, Hepato cellular carcinoma, Lupus anticoagulants. 2. Secondary cause of Budd Chiari Syndrome (which contributes to 25 %) includes Compression of the hepatic vein due to an abnormal growth (e.g.: tumor).(2),(3) The blood clot formation within the veins of liver may lead to occurrence of Budd Chiari Syndrome. This ailment can be sudden in onset and severe, acute or chronic in phase, or even asymptomatic. Sub acute penetration is the typical form mostly seen. Budd Chiari Syndrome also known as Hepatic venous thrombosis is secondary to cancer.(4)

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Incidence rate

It is a rare case. About 1 case per million population is reported in the incidence.[3]

Objectives:

1. To gain more knowledge in depth regarding Budd Chiari syndrome.

2. To study medical, surgical and nursing management of Budd Chiari syndrome.

Patient information

The patient was 46 years old. Her marital status is married, her education is 10th class and occupation is house wife. Her family income is 25,000/- month. She was admitted in A.V.B.R.H. in Cathlab ICU on 29/11/19.she was diagnosed as Budd Chiari Syndrome after undergoing investigations such as venography.

Patient had chief complaint of tenderness in abdomen along with fever, rigor and pain since 1 month.

Medical history: Patient is a known case of hypertension since 7 years for which she was taking medication Tab. Metaprolol 25 mg, patient also had an episode of cerebrovascular accident 7 years back and is taking medication Tab. Ecosprin AV 75 mg. it is doubted that the patient may be a case of cirrhosis of liver which is leading to Budd Chiari Syndrome.

Family history: There is no hereditary or genetic disorders in family.

Psychosocial history: Patient maintains good interpersonal relationship with family, friends and relatives.

Causes of BCS includes the following:

· The primary etiology of Budd Chiari syndrome was present in patient which results in approximately (75%) of blood clot of the veins of the liver. Blood clot of the hepatic vein is connected with the following conditions:

1. Polycythemia vera (type of a blood cancer, which makes the bone marrow to form red blood cells in excess)

2. Hepatocellular carcinoma (carcinoma of liver that begins with hepatocytes)

3. Pregnancy

4. Postpartum state

5. Oral contraceptives use

6. Paroxysmal nocturnal hemoglobinuria

7. Lupus anticoagulants

Clinical findings

Patient had tenderness in abdomen on palpation, yellowish discoloration of skin and whites of eyes. Hepatomegaly, spleenomegaly, ascites, body temperature was 100◦ F no other abnormalities was found on examination.

Diagnostic assessment

In kidney function test, Sr. Urea was increased that was 49 mg/dl than the normal range i.e. 7-17mg/dl.

CECT Abdomen: Atrophic and heterogenous liver with gross caudate lobe hypertrophy significant of cirrhosis of liver with non visualization of right and middle hepatic vein? Budd Chiari Syndrome. Spleenomegaly with multiple venous collaterals. In case, the results of liver function tests are abnormal, an imaging studies, typically Doppler ultrasonography is done. If results are unclear magnetic resonance imaging (MRI) of the blood vessels or Computed Tomography scan is done.

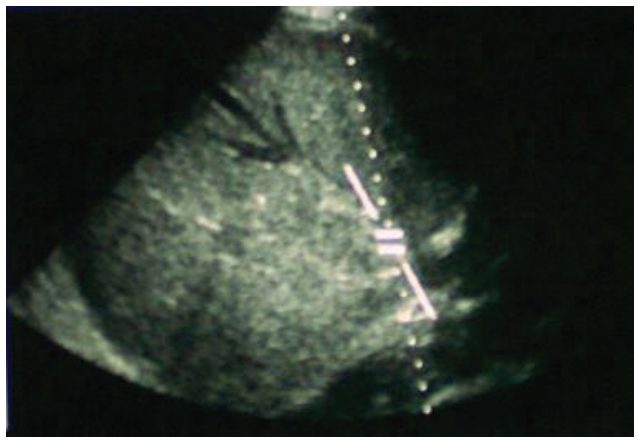

Fig1: Sonography showing hepatic vein thrombus.

Diagnosis: Diagnosis is made most commonly using retrograde angiography and ultrasound studies

of abdomen. Ultrasound studies may show obliteration of veins of the liver, blood clots or stenosis (narrowing of vessels), spider nevus, large abnormal vessels connecting aorta with pulmonary arteries, or a biliary atresia replacing a normal vein.

Therapeutic interventions

Collaborative management: A few numbers of patients can be treated pharmacologically with sodium restricted diet.

Pharmacologic management includes diuretics to control ascites, anticoagulants such as heparin and warfarin may also be prescribed depending on the condition of patient. Tab. Augmentin 625 mg (TDS, orally), Tab. Pantop 40mg (BD, orally), Tab. Ultracet 50mg (TDS, orally), Tab. Mucomix 600mg (orally) was prescribed for the patient.

Surgical management includes Transjugular Intrahepatic Portosystemic Shubt (TIPS) in milder cases which can divert the flow of blood around the obstruction of the liver. Liver transplantation in case of fulminant liver failure. Angioplasty (Baloon Cavoplasty to benefit from obstruction) was planned for this patient.

Nursing Management

1. Administer IV fluids as per doctors order
2. Administer medications as per doctors order
3. Maintain I/O chart
4. Monitor vital signs especially blood pressure as portal hypertension is also a major cause of Budd Chiari syndrome.
5. Providing patient and relatives with psychological support.
6. Encouraging patient to express emotions.
7. Establishing good interpersonal relationship.
8. Teaching coping strategies to the patient and relatives.
9. Maintaining confidentiality as it will relive the anxiety of the patient and relatives. It will help in avoiding the conflict with physicians, nurse and other health care providers.
10. Explain about surgical procedure to parents.
11. Explain about the prognosis of disease.

Follow up and outcomes: Patient was planned for follow up regularly on basis of advice given by physician.

Limitations: As this is a rare case conducting research studies on causes and treatment is difficult as the number of participants may be very less or rare.

Strength: This case is helpful to provide knowledge about thrombosis of hepatic vein especially in patients suffering from cirrhosis of liver.

Prognosis: The prognosis in patients suffering from BCS is very poor who especially in those who remains uncured without proper management, with death caused by progressive liver failure between the time duration of 3 months to 3 years since the time of the diagnosis.[4]

Discussion

The BCS also known as thrombus of veins of liver is a congestive hepatopathy which is caused due to blockage of the veins of liver. This condition is very sparse and is seen especially among the general population within 1/100,000. Hypercoagulable condition can be found in 75 percent of the patients; more than one causative factor may play a major role in 25 percent of the patients. [5]

The diagnosis is usually made using radio imaging techniques such as, (Computed tomography, or magnetic resonance imaging or Doppler ultrasound). Hepatic biopsy can be performed if Budd Chiari syndrome is suspected in small vessels. Management includes use of anticoagulation, the treatment of known risk factors for prothrombotic, percutaneous revascularization and transjugular intrahepatic portosystemic stent shunt to restore hepatic venous drainage, and liver transplantation in patients who does not respond. [6],[7]

The severity of the condition differs from case to case, depending upon the veins infected and their site. In few cases, if the major veins in liver are involved, high blood pressure may be present in the veins which carries blood from the gastrointestinal tract to the heart through the liver which is known as portal hypertension.

[8],[9] BCS produces mild to fatal liver damage; which differs from person to person.[10]

Informed consent: The patient and relatives were informed before taking the case.

Conclusion

46 years old female was admitted to AVBRH in Cathlab ICU with chief complaints of pain in abdomen, fever, rigor etc. after undergoing investigations she was diagnosed as Budd Chiari Syndrome. Budd Chiari Syndrome also known as Thrombus of the Hepatic Vein is a rare disorder characterized by narrowing and obstruction of the veins of the liver (hepatic veins).

Ethical Clearance: Author herself is a registered nurse and midwife (Completed Basic BSc Nursing and registered in Maharashtra Nursing hence professionally allowed to provide nursing care. During the authors clinical posting this client was allotted to her for providing nursing care.

Source of Funding: Self.

Conflict of Interest: NIL

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