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RESEARCH ARTICLE

RARE CASE OF BUDD-CHIARI SYNDROME (BCS) WITH RIGHT INGUINAL HERNIA: A CASE REPORT

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Abstract

Budd-Chiari syndrome (BCS) is an uncommon disorder characterized by obstruction of hepatic venous outflow. The obstruction may be thrombotic or non-thrombotic anywhere along the venous course from the hepatic venules to junction of the inferior vena cava (IVC) to the right atrium. Hepatic veno-occlusive disease and cardiac disorders are excluded from this definition. BCS is a heterogeneous clinical condition-it may be curable or potentially lethal.

Introduction:-

Budd-Chiari syndrome (BCS) is an uncommon disorder characterized by obstruction of hepatic venous outflow. The obstruction may be thrombotic or non-thrombotic anywhere along the venous course from the hepatic venules to junction of the inferior vena cava (IVC) to the right atrium. Hepatic veno-occlusive disease and cardiac disorders are excluded from this definition. BCS is a heterogeneous clinical condition-it may be curable or potentially lethal. (1)

Case Presentation:-

18 years old male patient, was diagnosed by abdominal ultrasound trans abdominal pre peritoneal hernia repair (TAPP).

Laparoscopy was done during inguinal hernia repair, ascites with liver congestion was noticed although the patient was very healthy before surgery and preoperative blood tests were normal.

Ascitic fluid sample was taken for cytology and microbiology.

As there is a risk of mesh infection- plan is changed from laparoscopic to open inguinal hernia repair.

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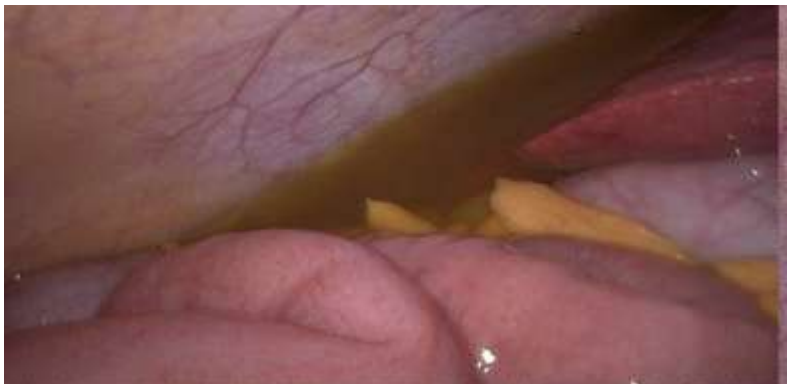

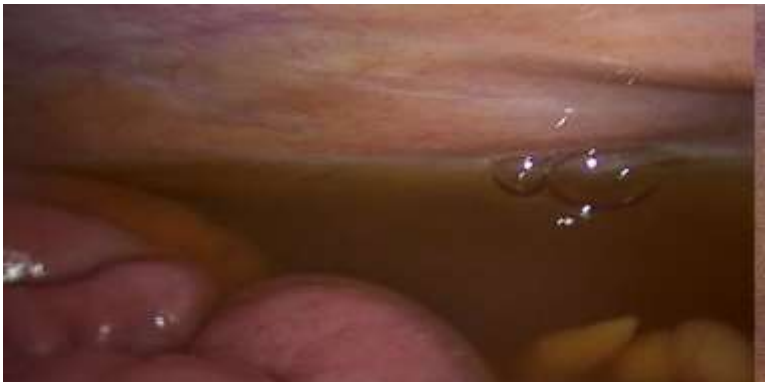

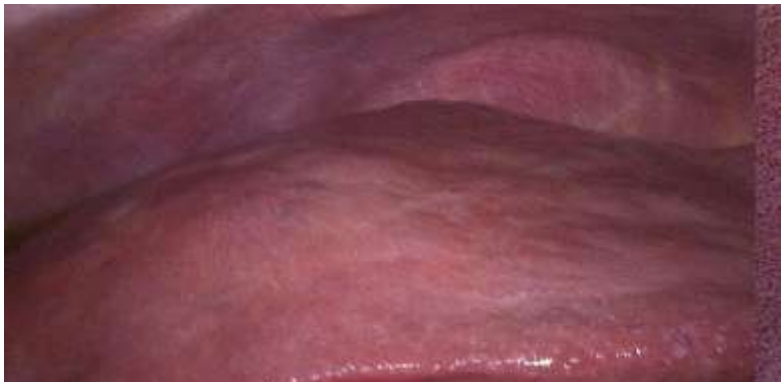

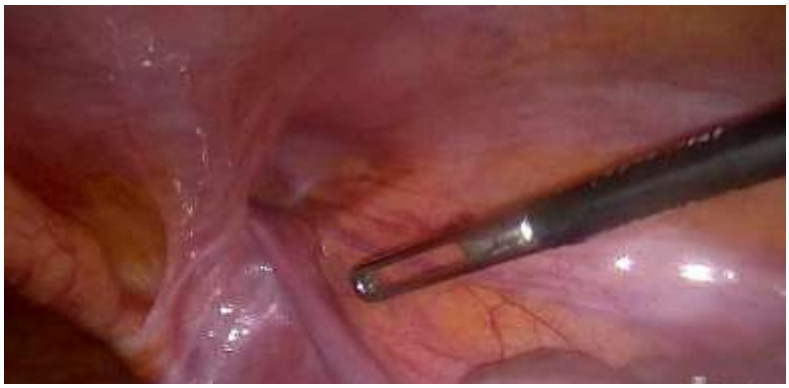

Postoperative abdominal ultrasound, CT scan and liver functions, hepatitis markers were done, patient was referred to gastroenterologist and cardiologist, diagnosed as BUDD CHARI SYNDROME. Anticoagulants, Colchicine,

Spironolactone, Ursodeoxy Cholic Acid, Furosemide are given, patient was improved clinically and his investigations were back to normal during 4 months.

Discussion: -

- **Etiology**

In 80 percent of the cases, some underlying cause leads to the development of Budd-Chiari syndrome, and the majority are related to a hypercoagulable state. This aspect of Budd-Chiari syndrome must be considered for diagnosis and treatment.**(1)**. The most important causes of Budd-Chiari syndrome are the following:

I. **Myeloproliferative Disorders**

Almost half of the cases of Budd-Chiari syndrome are related to some type of myeloproliferative disorder like polycythemia vera and essential thrombocythemia because these disorders are almost always accompanied by some type of hypercoagulability.

II. **Malignancy**

10% of Budd-Chiari syndrome cases are related to malignancy, which causes either direct compression or invasion of vessels. These, along with hypercoagulability, lead to venous thrombosis and obstruction. The most common cancer related to the Budd-Chiari syndrome is hepatocellular carcinoma, followed by cancers of the adrenal gland, renal cell carcinoma, leiomyosarcoma, right atrial myxoma, and Wilms tumor.

III. **Lesions of the Liver**

Sometimes, infection or a space-occupying lesion of the liver leads to the compression of the vasculature. Hepatic cysts, adenomas, cystadenomas, invasive aspergillosis, and aortic aneurysm are common lesions that lead to Budd-Chiari syndrome.

IV. **Pregnancy and OCPs (oral contraceptives)**

Oral contraceptives and pregnancy lead to a hypercoagulable state and are responsible for about 20 percent of cases of Budd-Chiari syndrome.

V. **Idiopathic**

20% of the cases are idiopathic.

Other hypercoagulable states responsible for Budd-Chiari syndrome include:

Factor V (Leiden) mutation that leads to protein C resistanceAntiphospholipid antibody syndromeAntithrombindeficiencyProtein C deficiencyParoxysmal nocturnal hemoglobinuria. **(2)**

- **Pathogenesis**

Blockage of two or more major hepatic veins increases the sinusoidal pressure and reduces sinusoidal blood flow. Obstruction of a single hepatic vein is generally not evident; two veins must be blocked for clinical disease. The result of these hemodynamic changes is sinusoidal dilation and filtration of interstitial fluid. Filtrated interstitial fluid passes through the liver capsule when it exceeds the capacity of lymphatic drainage **(3,4).**

- **Clinical presentation:**

- **Acute and subacute presentation:**

Sudden onset of ascites, abdominal pain, jaundice, hepatomegaly, renal failure, hepatic encephalopathy.

- **Chronic presentation (most common):**

Progressive ascites and absence of jaundice. Half of the patients can present with renal impairment.

- **Rare presentation:**

Tender hepatomegaly, fulminant/sub-fulminant hepatic failure, jaundice, and renal failure.

BCS should be suspected when there is:

1. Sudden onset of ascites and painful hepatomegaly
2. Massive ascites with relatively normal liver functions
3. Sinusoidal dilation in liver biopsy without heart disease
4. Fulminant hepatic failure along with hepatomegaly and ascites
5. Unexplained chronic liver disease
6. Liver disease with a thrombogenic disorder.**(5)**.

- **Diagnosis**

BCS should be suspected in patients with:
(1) Abrupt onset of ascites and painful hepatomegaly.
(2) Massive ascites with relatively preserved liver functions.
(3) Sinusoidal dilation in liver biopsy without heart disease.
(4) Fulminant hepatic failure associated with hepatomegaly and ascites.
(5) Unexplained chronic liver disease.
(6) Liver disease with thrombogenic disorder.

Serum transferase levels may be more than five times the upper limit of the normal range, especially in the fulminant and acute forms of BCS. Serum alkaline phosphatase and bilirubin levels also increase. Serum albumin level decreases moderately.

Doppler ultrasonography of the liver, with a sensitivity and specificity of 85% or more, is the technique of choice for initial investigation when BCS is suspected **(6)**.

Magnetic resonance imaging (MRI) should be performed as a second-line imaging modality. MRI can show the hepatic vein thrombosis and evaluate the IVC, but it is more expensive than computed tomographic (CT) scanning. **(7).**

Hepatic venography is the reference procedure for the evaluation of hepatic veins, extent of thrombosis and caval pressures. Inferior cavography should be performed to demonstrate stenosis or occlusion of the IVC.

Liver biopsy shows congestion, liver cell loss and fibrosis predominantly located in the centrilobular area. **(8).**

Conclusion: -

Budd-Chiari syndrome (BCS) is an uncommon condition characterized by obstruction of the hepatic venous outflow tract. Presentation may vary from a completely asymptomatic condition to fulminant liver failure. BCS is an example of postsinusoidal portal hypertension. The management can be divided into three main categories: medical, surgical, and endovascular. BCS requires accurate, prompt diagnosis and aggressive therapy. Treatment will vary depending on the clinical presentation, cause, and anatomic location of the problem. Patients with BCS are probably best treated in tertiary care centers where liver transplantation is available.

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