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Review Article

BUDD CHIARI SYNDROME-REVIEW ARTICLE

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A R T I C L E I N F O

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A B S T R A C T

Budd Chiari syndrome is a condition caused by occlusion of hepatic veins that drain the liver. Is a hetrogenous group of disoders the clinical variant of Budd-Chiari syndrome-acute and sub acute chronic fulminant the cause primary Budd-Chiari syndrome (75%) secondary Budd-Chiari syndrome (25%) diagnosis laboratory studies imaging studies imaging studies a minority of patient can be treated medically with sodium restriction, diuretics to control ascites pharmacologic therapy procedure and surgery in general 2/3 of patient with Budd-Chiari are alive

INTRODUCTION

Budd Chiari Syndrome Is A Hetrogenous Group of Disoders characterised by hepatic venous outflow obstruction at the level of the hepatic venules, the large hepatic veins, the inferior vena cava, or the right atrium. Hepatic veno-occulsion disease refers to obstruction of hepatic venous outflow at level of the central or sub lobular hepatic veins, or both

Signs and symptoms

- Physical examination will reveal the following
- Jaundice
- Ascites
- Hepatomegaly
- Spienomegaly
- Ankle edema
- Stasis ulcerations
- Prominence of collateral veins

The clinical variant of budd-chiari syndrome

Acute and sub acute- characterized by rapid development of abdominal pain, ascites (which can cause abdominal distention) hepatomegaly, jaundice, renal failure

Chronic form- most common presentation; patient present with progressive ascites; jaundice is absent; approximately 50% of patient also have renal impairment

Fulminant form- uncommon presentation fulminant or subfulminanthepatic failure is present, along with ascites, tender hepatomegaly, jaundice and renal failure

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Etiology

The cause can be found in more than 80% of the patient

Primary Budd-Chiari Syndrome (75%)

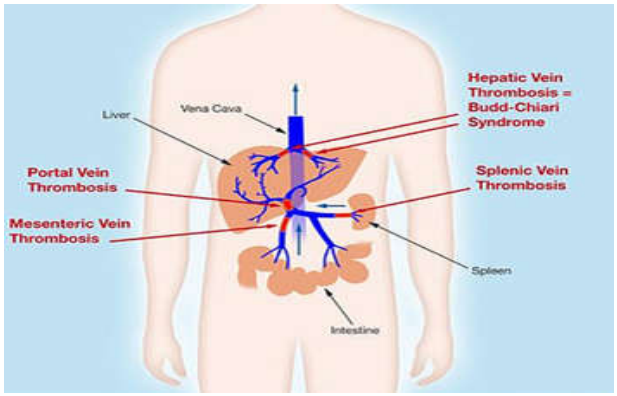
- Thrombosis of hepatic vein
- Hepatic vein thrombosis is associated with the following in decreasing order of frequency
- Polycythemiavera
- Pregnancy
- Postpartum state
- Use of oral contraceptives
- Paraoxysmal nocturnal hemoglobinuria
- Hepatocellular carcinoma
- Lupus anticoagulants

Secondary Budd-Chiari Syndrome (25%)

Compression of the hepatic vein by an outside structure (e.g a tumor) Budd-chiari syndrome is also seen in infection such as tuberculosis, congenital venous webs and occasionally in inferior vena caval stenosis

Pathophysiology

Any obstruction of the venous vasculature of the liver is referred to as budd-chiari syndrome, from the venules to the right atrium. This lead to increased portal pressure causes increased filtration of vascular fluid with formation of ascites in the abdomen and collateral venous flow through alternative veins leading to esophageal, gastric and rectal varices. Obstruction also cause centrilobular necrosis and peripheral lobule fatty changes due to ischemia. If the condition persists nutmeg liver will develop.



Renal failure may occur, due subsequent activation of rennin angiotensis pathways and excess sodium retention

Diagnosis

Laboratory studies

- Examination of ascetic fluid provide useful clues to thwe diagnosis odbuddchiari syndrome,
- Patient usually have high protein concentrations (>2g/dl); this may not be present in person with acute form of the disease
- The white blood cell (WBC) count is usually less than 500/ml
- The serum ascites- albumijn gradient is usually less than 1.1(except in the acute forms of budd-chiari syndrome

Imaging Studies

- ultrasonography
- computed tomography
- magnetic resonance imaging (MRI)
- Venography

Biopsy

Pathological finding in liver biopsy are
High grade venous congestion and centrilobular liver cell atrophy

Thrombin within the terminal hepatic venules

Management

A minority of patient can be treated medically with sodium restriction, Diuretics to control ascites

Pharmacologic therapy

- Anticoagulants
- Thrombolytics
- Diuretics

Procedure and surgery

- Balloon angioplasty
- Localized thrombolysis
- Placement of stent or transjugular intrahepatic portacaval shunt (TIPS)
- Variceal treatment
- Paracentesis
- Portal decompression
- Precutaneoustranshepatic balloon angioplasty (PTBA)
- Liver transplantation

Prognosis

When the vein remain completely blocked, most people, if untreated, die of liver failure within 3 years. When blockage is incomplete, life expectancy is longer but varies. In general 2/3 of patient with budd-Chiari are alive at 10 years important negative prognostic indicators include ascites, encephalopathy, elevated child-pugh scores, elevated prothrombin time and altered serum level of various substance (sodium, creatinine, albumin, and bilirubin). Survival is also highly dependent on the underlying cause of the budd-Chiari syndrome.

CONCULSION

To conclude Budd chiari syndrome is a condition caused by occlusion of hepatic veins that drain the liver. It present with classical triad of abdominal pain, ascites and liver enlargement. The formation of a blood clot within the hepatic veins can lead to budd-chiari syndrome. It occus in 1 out of million individuals. The syndrome can be fulminate acute or symptomatic

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