Budd-Chiari Syndrome

Budd-Chiari syndrome is a rare hepatic disorder of obstructed venous outflow from the hepatic veins. Patients develop hepatomegaly with centrilobular congestion and necrosis of the liver. Patients can develop ascites, abdominal pain, and on gross pathology have a "nutmeg liver." Causes for this syndrome include tumor, chronic infection, hematologic disorders, along with pregnancy and postpartum changes.



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Causes

Pregnancy and Postpartum

Pregnant-woman and Post-baby

Budd-Chiari syndrome is also associated with pregnancy, along with the postpartum state in females.

Hematologic Disorders

He-man-blood Disordered

Hematologic disorders such as polycythemia vera, paroxysmal nocturnal hemoglobinuria, antiphospholipid antibody syndrome, and essential thrombocytosis are possible causes. Other inherited coagulopathies include the following: Protein C deficiency, Protein S deficiency, Antithrombin III deficiency and Factor V Leiden. Hypercoagulability can lead to hepatic venous thrombosis.

Tumors

Tumors

Tumors or metastatic disease; specifically hepatocellular carcinoma (HCC), renal cell carcinoma, leiomyosarcoma, adrenal carcinoma, Wilms' tumor and right atrial myxoma are associated with Budd-Chiari. Tumors can cause hepatic venous outflow obstruction via mechanical obstruction and compression of the IVC. Furthermore, cancer can lead to a hypercoagulable state, which can also lead to a predisposition for thrombosis.

Chronic Infections

Crone with Infectious bacteria and viruses

Chronic infections can predispose patients to developing Budd-Chiari syndrome. Specific examples include hydatid cysts, aspergillosis, amebic abscesses, syphilis, and tuberculosis.

Mechanism

Thrombosis of Major Hepatic Veins

Trombones in Major Hepatic Vein

The most common pathophysiology for this syndrome is thrombosis of the major hepatic veins, preventing venous flow from leaving the liver. This does not always have to be a thrombotic cause, and mechanical obstruction can also lead to this syndrome.

Centrilobular Congestion and Necrosis

Centrilobular Congested-traffic and Necrosis-crows

When venous flow of the liver is compromised, the liver becomes enlarged and the sinusoids are engorged with blood. There is centrilobular congestion, and decreased oxygen transport leads to tissue necrosis.

Symptoms

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Abdominal Pain

Abdominal Pain-bolt

Patients can complain of abdominal pain with this disorder.

Ascites

Ascites Iced-tea

Ascites is a common feature of Budd-Chiari syndrome. This occurs because of two different pathophysiologies. As venous flow is backed up, lower extremity edema forms. Later in the course of disease, when liver damage occurs, albumin production decreases. This decreases vascular osmotic pressure, allowing fluids to third-space into tissues.

Hepatomegaly

Liver-balloon

Patients display hepatomegaly on exam with Budd-Chiari syndrome, and elevated liver enzymes may be seen. This may correlate to other hepatic disorders, like liver failure, encephalopathy.

Nutmeg Liver

Nutmeg

"Nutmeg liver" is a description of the gross appearance of the liver with chronic Budd-Chiari syndrome. This term describes the mottled, dark red congested regions that represent accumulation of RBC's in centrilobular regions on gross pathology.

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