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the irreplaceable image

Budd-Chiari syndrome in chronic myeloid leukemia

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A 29-year-old Ph-positive patient in accelerated phase chronic myeloid leukemia (CML) with marked thrombocytosis developed fever, abdominal pain and signs of severe acute hepatitis, although was negative for viral markers. Real-time and Doppler ultrasound (US) scan showed marked hepatomegaly, caudate lobe hypertrophy (Figure 1), failure to visualize hepatic veins and their flow, portal vein ectasy with slow hepatopetal flow, splenomegaly and massive ascites. MRI and angiography confirmed occlusion of all hepatic veins and partial obstruction of the inferior vena cava hepatic segment, likely due to disproportionate caudate lobe enlargement. These findings suggested Budd-Chiari syndrome (BCS); treatment was diuretics, anticoagulants, chemotherapy and peritoneum-jugular shunt. Liver histology, obtained by uncomplicated percutaneous biopsy, confirmed the diagnosis (Figure 2). Five months later the patient is still in accelerated phase, with normal platelet count, moderate liver dysfunction and unmodified US abnormalities. Surgical decompression by portacaval shunt is now considered. BCS may induce acute liver failure in CML patients. Liver US monitoring is essential in patients with myeloproliferative syndromes at risk of thrombosis.

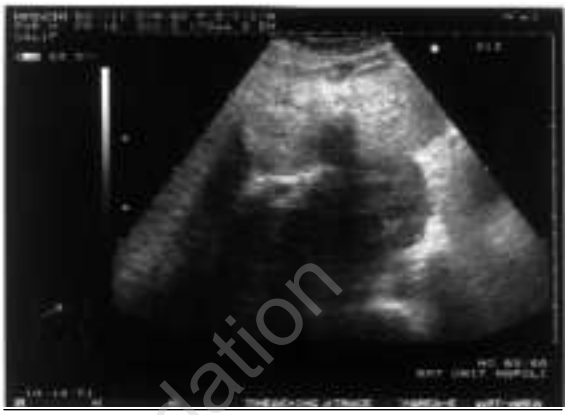

Figure 1. Ultrasound scan of liver showing caudate lobe hypertrophy, a typical finding in BCS.

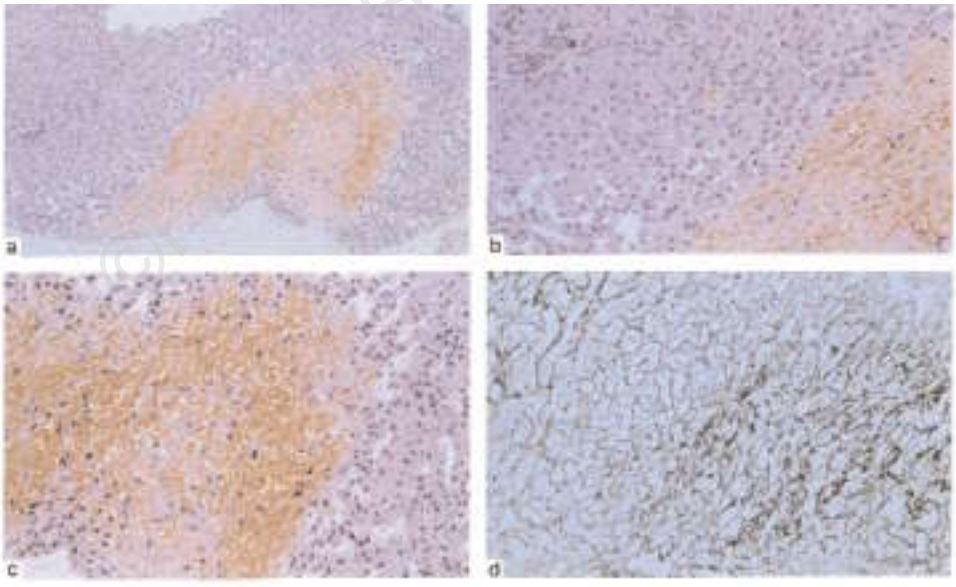

Figure 2. Histologic findings: centrolobular hemorrhage (a); higher magnification, showing a normal portal area (b) and hepatocyte necrosis (c); reticular collapse (Gomori staining) (d).

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