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Budd-Chiari Syndrome: Common Presentation, Rare Disease

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Abstract : Background: Budd-Chiari syndrome is caused by thrombosis of the hepatic veins and/or the thrombosis of the intrahepatic or suprahepatic IVC. The etiology remains idiopathic in 16% -35% of cases. Malignancy, rheumatological disorder, myeloproliferative disease, inheritable coagulopathy, infection or hyperestrogen state can be identified in many cases. Methodology: Review of case records of the patient presented to Aster Medcity, Emergency Department, Cochin. Introduction:17 years old female was presented to ED with fever, jaundice and abdominal distention since 1 week. O/E: Pallor+, icterus+. Abdomen- gross distension+, shifting dullness+, generalized anasarca+. USG abdomen showed hepatomegaly with mild coarse echotexture and moderate to gross ascites. CT abdomen and chest showed hepatomegaly with thrombosis of all three hepatic vein and moderate ascites suggestive of Budd-Chiari syndrome. Patient was taken for catheter vein thrombolysis. Venogram done the next day revealed almost > 50% opening of the right hepatic vein. Concurrent doppler showed colour and doppler signals in middle hepatic veins. She gradually improved and was discharged home on anticoagulant and adviced regular follow up. Conclusion: Being a rare disease in this young population, high suspicion is required when evaluating young patients with abdominal pain and jaundice.

Keywords : Budd-Chiari syndrome, rare disease, abdominal pain, India
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