A 28-YEAR –OLD MAN PRESENTED WITH CAROLIS SYNDROME WITH MASSIVE SPLENOMEGALY AND MULTISYSTEM INVOLVEMENT

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Carolis Syndrome is a rare congenital fibrocystic liver disease defined by the combination of Carolis Disease (cystic dilatation of intrahepatic bile ducts) and Hepatic Fibrosis (HF). This syndrome typically leads to non-cirrhotic portal hypertension. While it often presents in childhood, adult cases can manifest with severe hypersplenism and rare extrahepatic associations. A 28-year-old male presented with a 3-year history of a progressive abdominal lump. Recurrent low-grade fever, cough, and significant weight loss (~10 kg) for last 6 month. His physical examinaiton were Ill looking, Poor nutritional status, significant pallor and non-icterus, lymphadenopathy or stigmata of chronic liver disease Massive splenomegaly (18 cm below the left costal margin). No hepatomegaly or ascites and other systemic examination unremarkable Carolis syndrome, though rare, should be considered in young adults presenting with massive splenomegaly and portal hypertension. This case highlights that the disease can manifest as a complex multisystem disorder, involving hepatic, renal, and pulmonary systems as fibrocystic ciliopathies Early recognition and a multidisciplinary approach are essential for managing its diverse clinical sequelae and improving patient outcomes.

Keywords: *Carolis Syndrome, Massive Splenomegaly, Hepatic Fibrosis, Nephrocalcinosis, Bronchiectasis,Liver Elastography.*

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