ABERNATHY SYNDROME: AN UNUSUAL CAUSE OF PAIN ABDOMEN

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INTRODUCTION:

Abernathy syndrome, widely calledCongenital extra-hepatic portosystemic Shunt (CPES) is a rare condition with a prevalence of 1 in 30000 -50000 cases, where the portal venous system is diverted away from the liver into the systemic veins.

It is classified into (Type I) with absent portal vein in which there is no perfusion of liver by the portal vein and (Type II) in which few thin radicles of portal vein are present and portal blood gets partially shunted to the inferior vena cava.

Early diagnosis helps in preventing the harmful effects of bypassing of liver. Initially, patients present with symptoms note related to liver such as chronic/ recurrent abdominal pain and fatigue.

In later course of disease hepatic parenchymal abnormalities are seen including nodular liver lesions or tumors such as hepatoblastoma/ hepatocellular carcinoma and life-threatening conditions such as hepatopulmonary syndrome, hepatic encephalopathy.

BACKGROUND:

A13 year oldmale presented with complaints of recurrent attacks of abdominal pain and mild fatigue.The abdomen was soft on examination with no organomegaly or ascites. Routine blood investigations were normal with normal liver function tests.

Ultrasound showed a normal liver parenchyma with portal blood draining into the inferior vena cava, following which a CT angiogram was advised.

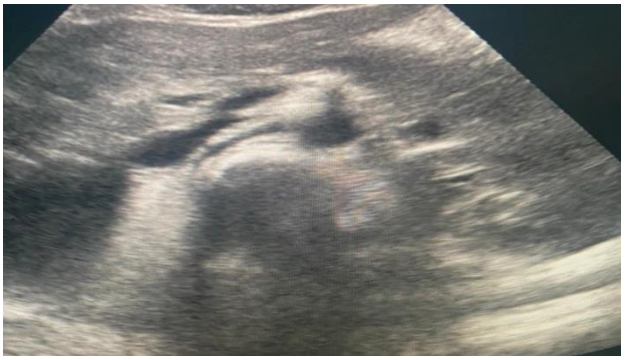

Fig 1: Ultrasound image showing a normal liver parenchyma with portal blood draining into the inferior vena cava.

CT angio with MRCP correlation revealed confluence ofSMV along with splenic vein and this common trunk wasseen draining into inferior vena cava with a dilated portal vein at its formation measuring~14mm and a small collateral from the portal vein was seen supplying the liver. The SMV and splenic vein common trunk was seen joining the inferior vena cava at the level of pancreatic head.

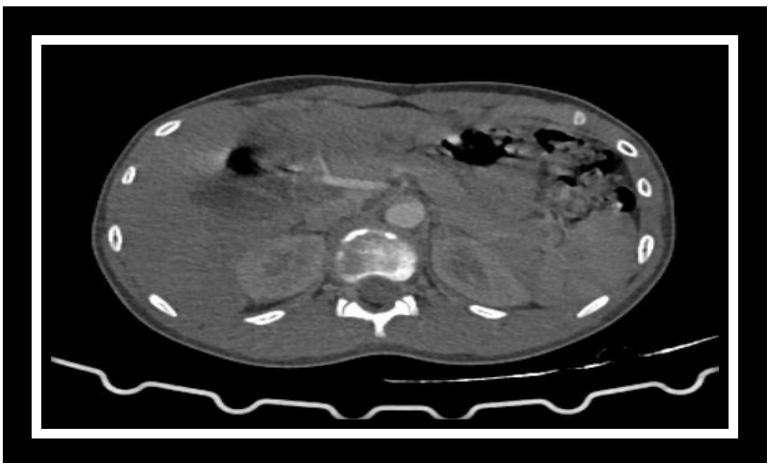

Fig 2 :Axial CT (Arterial phase) showing hepatic artery coursing towards the hilum.

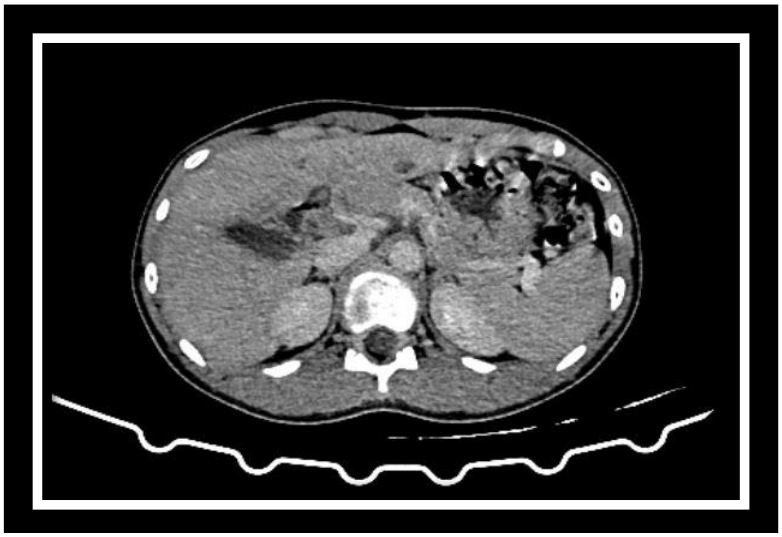

Fig 3: Axial CT image in venous phase showing small collateral vessel arising from the common trunk just before draining into IVC.

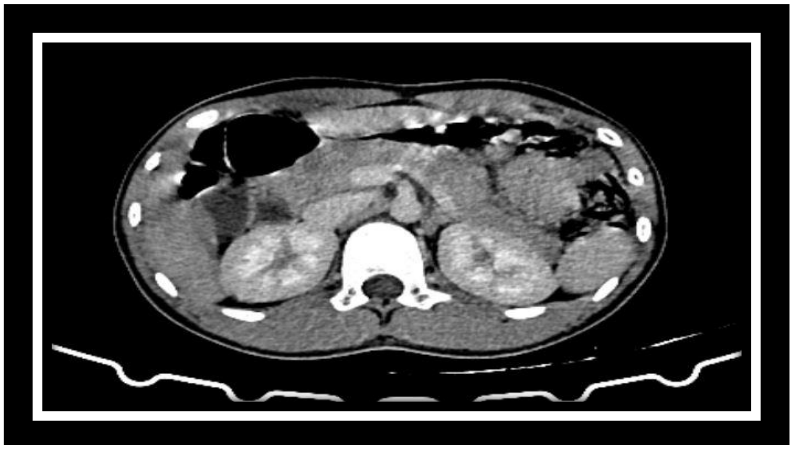

Fig 4: Axial CT (Venous phase) showing SMV and splenic vein joining to form a common trunk

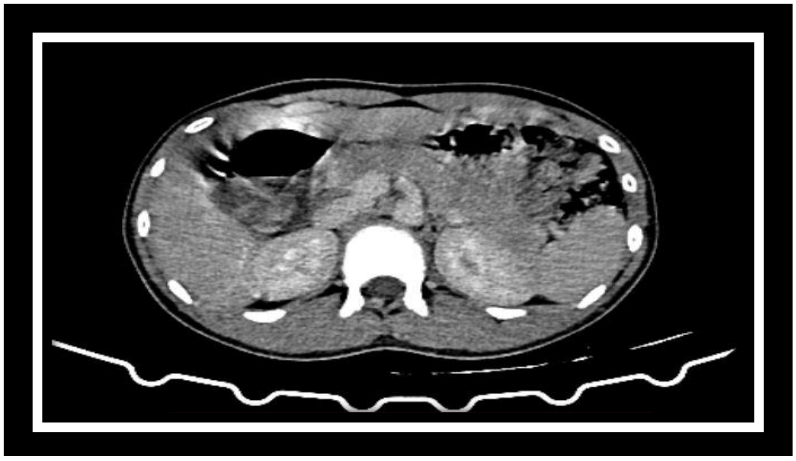

Fig 5: Axial CT (Venous phase) Common trunk (Black arrow) is seen draining IVC (white arrow

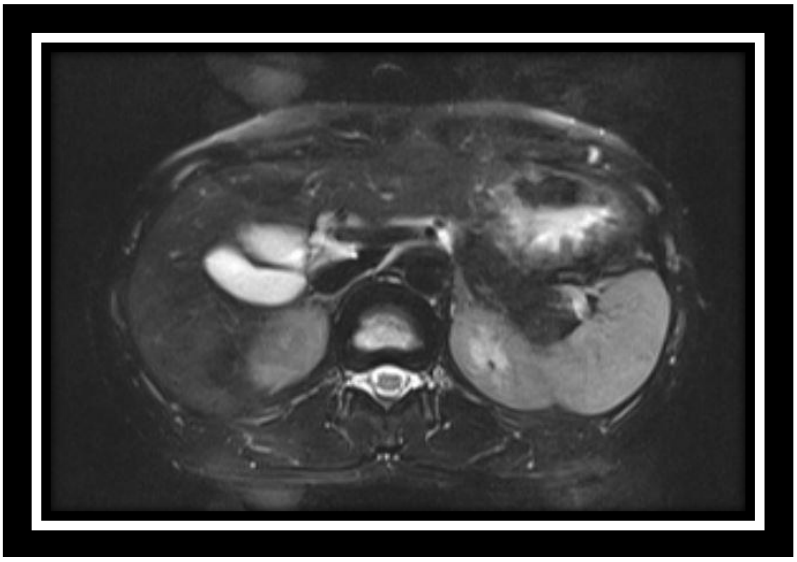

Fig 6: Axial T2 Turbo spin echo Fat suppressed image showing common trunk (White arrow) draining IVC (White block arrow).

Based on the above finding's patient was diagnosed with Type II Abernathy syndrome.Following which patient is now planned for shunt closure and reconstruction of the portal vein.

CONCLUSION:

Abernathy syndrome is a type of rare congenital vascular anomaly. Rarely the patients may present with indeterminate symptoms such as abdominal pain and hence go unrecognized. Thus, it should be a differential while evaluating unknown cause of pain abdomen for early detection and preventing harmful effects of bypassing of liver. CT protocol modification may be required for an accurate diagnosis.

Based on type of Abernathy malformation patients should be planned for surgery.

In Type I closure of the shunt isn't usually performed because it may be the only drainage of mesenteric veins.

In Type II shunt closure is performed along with reconstruction of portal vein to prevent the detrimental effects of bypassing liver. These patients require a long standing follow up.

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