

Courveilhier Baumgarten Syndrome: A Rare Syndrome Revisited

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Abstract: A 25 year old male had presented with history of abdominal distension of 20 days duration and on examination he had prominent abdominal vein and a venous hum with Ascites. He had no history of altered sensorium, melena. The clinical scenario was suggestive of Cruveilhier Baumgarten syndrome.

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Introduction

The term Cruveilhier Baumgarten syndrome is used for cases of portal hypertension due to any cause in which a loud venous murmur can be heard over the upper abdomen. The Cruveilhier Baumgarten disease is reserved for cases with congenital patency of the umbilical vein associated with congenital hypoplasia of the liver and portal system and a venous murmur heard over the umbilical vein⁽¹⁾.

Case Scenario

25 year old male was admitted with history of progressive abdominal distension of 20 days duration and fever of 4 days duration. Patient had off and on abdominal distension for last 3 years and no history of altered sensorium or hematemesis. This illness had affected his schooling and he had discontinued his studies. He was 3rd in birth order and had an uneventful birth history, normal developmental history and an unremarkable family history. On examination patient was conscious oriented with pallor. no lymphadenopathy, jaundice or clubbing.

Patient had sparse axillary and pubic hair. He was hemodynamically stable. His abdominal examination revealed a prominent distended vein on the abdomen. The flow of blood in this vein was towards umbilicus and there was no umbilical hernia (Fig.1)



Fig. (1). Prominent Vein near Umbilicus.

No palpable thrill was appreciated. However a loud venous hum was heard on auscultation. He also had moderate splenomegaly and gross ascites. His other systemic examination was unremarkable. On evaluation patient had hemoglobin of 7gm/dl (normal 12-14gm/dl), TLC of 3500 (normal 4500-10,000), platelet count of 60,000 (normal 150,000-400,000). PBF was normocytic

and normochromic and his corrected retic count was 3%. He had serum Bilirubin levels of 1.2 mg/dl. He had normal (SGOT/SGPT 37/43 IU). However, his serum albumin levels were 2.9mg/dl (normal 3.5gm/dl). PT/INR was deranged 45 sec/16 sec, INR 3.5. Ascitic fluid analysis showed high SAAG 1.8 with predominance of neutrophils with absolute neutrophil count > 360/mm². Upper GI endoscopy showed two columns of Grade 1 esophageal varices with no features of congestive gastropathy or fundal varices. Ultrasound abdomen showed main portal vein and intrahepatic branches of PV are dilated (fig.2 and 3). Collaterals are seen in ligamentum teres and periumbilical region, representing recanalized peri-umbilical veins (Fig.2-4).

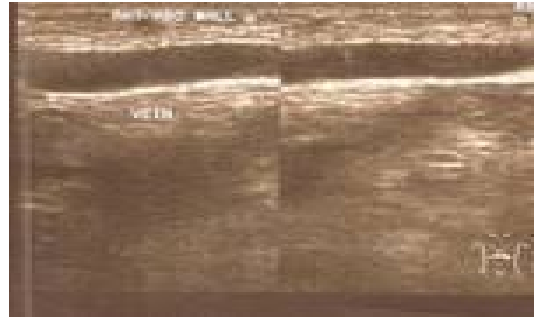


Fig. (2). USG showing patent abdominal vein: CBS.

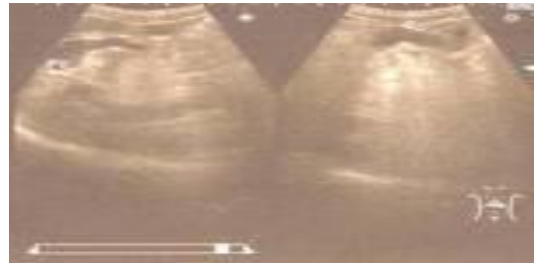


Fig. (3). USG showing Dilated Portal Vein.



Fig. (4). USG showing Dilated Portosplenic axis suggestive of portal hypertension with features of chronic liver disease.

Liver span was reduced with coarsening of echotexture with enlarged caudate lobe and moderate splenomegaly and ascites. His viral markers, auto immune profile, Wilson's profile, iron profile was all negative. *The initial diagnosis* was chronic liver disease non-bleeder (cryptogenic), child Pugh score 9, (class-B) portal hypertension, spontaneous bacterial peritonitis, and *Courveilhier Baumgarten Syndrome*. He was put started on antibiotics and diuretics besides other supportive care. His fever settled and had decrease in ascites. Patient is doing well and is following our Out patient department.

Discussion

In 1833 Pegot reported a case of portal hypertention in which a loud venous hum was heard at the umbilicus. This case was elaborated by cruveilhier. At autopsy it was found that the venous murmur was due to collateral circulation through a widely patent umbilical vein. The liver was hypoplastic. In 1908 Baumgarten reported a similar case in a 16 year old boy who died following a gastric haemorrhage. He believed that the widely patent umbilical vein, splenomegaly and atrophic liver were due to congenital hypoplasia of the liver. Since then several similar cases have been reported in foreign literature but the first complete review in the English literature was that by Neil Armstrong et al² in 1942. The portal circulation commonly decompresses through collaterals in the ligamentum teres. This usually echogenic structure becomes sonolucent centrally, producing a "bull's eye" appearance in the transverse plane on ultrasound examination. A central vascular channel exceeding 3mm in diameter is a specific sign of portal hypertension. On longitudinal scans, these recanalized paraumbilical veins can be followed caudally toward the umbilicus as a tubular lucency (Fig 3). A patent umbilical vein

excludes an extra hepatic cause of portal hypertension because the umbilical vein arises from the intrahepatic portion of the left portal vein. This vein enables the formation of an anastomosis between the left branch of the portal vein and the veins of the anterior abdominal wall, creating a portal systemic bypass circuit known as the Cruveilhier Baumgarten syndrome. The vein may sometimes become aneurysm ally dilated and simulates a pancreatic pseudocyst, so Doppler scanning of cystic structures in patients with cirrhosis should be done before Biopsy. Doppler sonography can also be used to assess the hemodynamic significance of flow in paraumbilical vein. When hepatofugal flow in the umbilical vein exceeds hepatopetal flow in the portal vein, patients are less likely to have esophageal varices and bleeding. Although it occurs more commonly in patients with severe functional impairment, it may play a protective role against variceal bleeding.^(3,4,5)

Our case emphasizes the importance of realizing this rare case in surgical practice.

References

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