A 25-year-old woman came to the hospital with the feeling of a lump in the left hypochondrium for one year. On examination, she was found to have massive splenomegaly, mild pallor, icterus and dilated veins in the infraumbilical region over which a venous hum was present. Laboratory examination confirmed her anti-HCV reactivity. The aspartate transaminase, alanine transaminase levels were 57.97 and 34.99 IU/l respectively. She had serum alkaline phosphatase of 104 kAU; total protein was 6.59 gm/dl and albumin was 3.5 gm/dl. There was no history of hematemesis, somnolence or mental confusion. Gray scale and Doppler ultrasound abdomen revealed coarsened liver echotexture with nodular outline suggestive of chronic liver parenchymal disease. The main portal vein and its right as well as left branches were dilated with hepatopetal flow in all segmental portal veins [Figure 1a]. There was gross splenomegaly and multiple collaterals were observed at the splenic hilum and anterior abdominal wall [Figure 1b]. The paraumbilical vein was patent and had hepatofugal flow, and was communicating with the anterior abdominal wall collaterals. No ascites was present. Multidetector computed tomography was performed on a 16-slice scanner (Sensation 16, Siemens Medical Solutions) after injecting 100 ml of non-ionic contrast at a rate of 4 ml/sec; using tube current of 200 mAs, tube voltage of 120 kVp and pitch of 1.5; with image acquisition in both arterial and venous phases. The axial images were post-processed on a workstation and supplemented by multi-planar reformations, volume rendering, and maximum intensity projections. The computed tomography (CT) confirmed heterogeneous nodular liver with dilated portal vein [Figure 2a], splenic enlargement and multiple collaterals in the subcutaneous plane on the anterior abdominal wall [Figure 2b,c]. The volume rendered images nicely depicted the infraumbilical bunch of collaterals connected to the left branch of the portal vein via a very large, tortuous paraumbilical vein [Figure 3a-c].

Cruveilhier Baumgarten (CB) syndrome comprises spontaneous portosystemic collateralization between the paraumbilical vein, the periumbilical veins of the anterior abdominal wall and the superficial and deep epigastric veins reaching the external iliac veins in a patient with portal hypertension. In classic CB syndrome, the umbilical portion of the left portal vein feeds a paraumbilical vein which leaves the liver and then heads towards the umbilicus. On CT the paraumbilical vein is seen as a tubular structure arising in the fatty falciform ligament between the left lobe of the liver, leading from the left portal vein to veins of the anterior abdominal wall. The network of dilated veins around the umbilicus gives a “Caput medusae” appearance.

Figure 1: (a) Gray scale sonogram showing heterogeneous liver echotexture and grossly dilated main portal vein. (b) Massive splenomegaly with intra-splenic echogenic foci and splenic hilar collaterals are also observed
The development of large recanalized paraumbilical vein has been found to prevent formation of bleeding esophageal varices and to predispose to hepatic encephalopathy. The clinical importance lies in the fact that these subcutaneous collaterals may undergo spontaneous hemorrhage or inadvertent significant bleeding during abdominal surgery or paracentesis. Multidetector computed tomography is a noninvasive method of diagnosis, which expeditiously evaluates the overall status of portosystemic collaterals in patients with portal hypertension.

References