

CRUVEILHIER BAUMGARTEN SYNDROME - RARE CAUSE OF UMBILICAL HERNIA IN LIVER CIRRHOSIS - ROLE OF RADIOLOGICAL IMAGING

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ABSTRACT

Umbilical hernia with dilated paraumbilical collateral vein as its content is rare. Diagnosing this condition is extremely crucial as rupture of the herniated umbilical vein during herniorrhaphy in a patient with ascites can be a fatal complication. Here we are reporting a case of a 37-year-old male who presented with complaints of abdominal distension, hematemesis, umbilical hernia with pulsations felt over the herniated sac. On ultrasonography, features of liver cirrhosis, portal hypertension and umbilical hernia with a herniated dilated vessel showing venous waveform on Doppler were seen. Contrast enhanced computed tomography (CECT) showed signs of liver parenchymal disease and dilated left paraumbilical vein coursing just beneath the anterior abdominal wall towards umbilicus with herniation through the umbilical hernia. Porto-systemic collaterals from this were then seen to be draining into right common femoral vein. Looking at the clinical and radiological features – diagnosis of Cruveilhier Baumgarten syndrome was observed.

Keywords: Cruveilhier Baumgarten Syndrome, Liver Cirrhosis

Case report

A 37-year-old male presented with hematemesis and melaena. There was no abdominal pain, fever, loose stools or urinary complaints. He was not a known case of diabetes or hypertension. He was a known alcoholic for 15 years. On examination - patient had abdominal distension and umbilical hernia which increased on straining with pulsations felt over the herniated sac. Clinically patient had jaundice with deranged liver function tests. On ultrasonography [Figures 1-3], liver showed coarse echotexture with irregular hepatic surface. Multiple collaterals were

noted at porta hepatis and splenic hilum. There was mild ascites and mild splenomegaly. At the site of umbilical hernia, a linear anechoic tubular structure was seen which showed mild pulsations. On color Doppler, it showed venous flow and waveform with a PSV of 33 cm/sec. Contrast enhanced computed tomography (CECT) of Abdomen & Pelvis [Figures 4-8] showed shrunken right hepatic lobe, nodular hepatic surface, caudate lobe hypertrophy with caudate to right lobe ratio of 81.9%. Hepatic artery and its branches appeared hypertrophied in arterial phase. Hepatic arterio-portal shunting was seen. Mild dilatation of portal vein (14 mm) at porta hepatitis was seen without thrombosis. Splenic vein measured 6.5 mm. Multiple dilated collaterals were noted at porta hepatis, peripancreatic region, splenic hilum, lieno-renal region, para-oesophageal region and in oesophageal wall. Left paraumbilical vein was dilated coursing just beneath the anterior abdominal wall towards umbilicus with herniation through the defect in linea alba which measured 15 mm in transverse and 8 mm in Craniocaudal extent. Dilated left para umbilical vein measured 15 mm in calibre. Dilated veins were noted in anterior abdominal wall coursing towards right common femoral vein (Porto-systemic collaterals). Splenomegaly was noted with splenic index of 1066. Splenic vein appeared tortuous. Ascites was noted. The diagnosis of chronic liver parenchymal disease-causing liver cirrhosis with portal hypertension, splenomegaly, ascites with multiple collaterals was made. Herniation of left para umbilical vein in umbilical region- Cruveilhier Baumgarten syndrome was observed.

Discussion

Umbilical hernias can be congenital and acquired. Congenital hernias develop in infancy and early childhood due to incomplete closure of the umbilical ring. They disappear spontaneously. Acquired hernias occur during adult life after the normal closure of the umbilical ring. ^[1]

Acquired hernias occur commonly in women after child birth especially multiple pregnancies. Weak abdominal wall, obesity, ascites, intra-abdominal masses and liver cirrhosis are predisposing factors. Strangulation of the intestine or omentum in the defect, incarceration causing intestinal obstruction and umbilical rupture caused by ascites are complications. Umbilical hernia with dilated paraumbilical collateral vein as its content is rare. It can be diagnosed on ultrasound with colour Doppler and CT with contrast. Ultrasound shows tubular, anechoic lesion within the umbilical hernia showing colour flow on colour Doppler and portal venous waveform on spectral Doppler. Contrast CT shows dilated left paraumbilical vein within the ligamentum teres coursing just beneath the anterior abdominal wall towards the umbilicus with herniation of the vein in umbilical region. ^[1,2]

Skin ulceration with subsequent rupture of the hernial sac can be a complication of umbilical hernia. ^[3] Diagnosing this condition is extremely crucial as rupture of umbilical hernia in a patient with ascites can be a fatal complication. Hence surgical repair is often recommended. Herniorrhaphy may damage the paraumbilical vein during surgery which can cause fatal haemorrhage. If varices are encountered during umbilical hernia repair, ligation of communicating vessels should be considered. ^[4] Hence preoperative recognition of the large paraumbilical vein in the surgical field is mandatory by colour Doppler to reduce the risk of torrential haemorrhage. CT abdomen with contrast in portal venous phase improves the diagnosis by tracing the dilated left paraumbilical vein from the liver to the umbilicus. Sometimes, opacified small bowel loops with oral contrast may have similar attenuation and hence be confused with enhancing vein. ^[5,6,7]

Portal hypertension causes collateral flow in the left paraumbilical vein which hence appears dilated. Collateral flow from left paraumbilical vein can reach systemic venous system by

various routes. Superiorly, these can drain into superior vena cava via lateral thoracic vein and axillary vein, and through superficial epigastric vein to the intra mammary vein and then subclavian vein. Inferiorly these can drain through circumflex iliac vein and superficial epigastric vein to the femoral vein and via inferior epigastric vein to the external iliac vein. ^[4]

Cruveilhier Baumgarten disease refers to congenital failure of closure of umbilical vein causing distension of paraumbilical vein. There is no liver disease usually found on imaging and liver biopsy. ^[8,9]

Cruveilhier Baumgarten sign refers to the venous hum heard at umbilicus due to caput medusae. It occurs due to hepatofugal flow through the recanalized umbilical vein.

Cruveilhier Baumgarten syndrome refers to the presence of a dilated and tortuous paraumbilical vein in the umbilical region in patients of liver cirrhosis. It arises from the left portal vein branch, traverses along the falciform ligament towards the umbilicus and forms a network of dilated paraumbilical veins. These eventually drain via superficial and deep epigastric veins into external iliac or femoral veins (systemic circulation). Awareness of this syndrome is must to avoid unintended iatrogenic injury of the vessels causing life threatening haemorrhage during paracentesis or abdominal surgeries. ^[7]

It was first described by Pegot in 1833 and then by Jean Cruveilhier in 1835 and Paul Clemens von Baumgarten in 1908. ^[8]

References

- 1) Khati NJ, Enquist EG, Javitt MC. Imaging of the umbilicus and periumbilical region. *RadioGraphics* 1998;18:413–431
- 2) Killi RM, Ozutemiz O, Elmas N. Color Doppler Sonography of Herniated Paraumbilical Collateral Vein Masquerading as an Acquired Umbilical Hernia; *Journal of Roentgenology*. 2000;174: 1465-1466. 10.2214/ajr.174.5.1741465
- 3) McAlister V. Management of umbilical hernia in patients with advanced liver disease. *Liver Transplantation* 2003;9:623– 625.
- 4) Hung TH, Hsiao FT, Tseng CW. Umbilical hernia due to enlarged paraumbilical vein. *Clinical Gastroenterology and Hepatology*. 2011 Sep 1;9(9):A30.
- 5) Sodhi JS, Zarger SA, Khan MA, Javid G, Khan BA, Shah AH, Mohd Gulzar G, Singh M (2007) Cruveilhier- Baumgarten syndrome revisited. *Indian J Gastroenterol* 26:173 (PMID: 17986745)
- 6) Sodhi KS, Saxena AK, Khandelwal N, Dhiman RK (2010) Giant paraumbilical veins in Cruveilhier-Baumgarten syndrome. *Gastrointest Endosc* 72:435-6 (PMID: 20538272)
- 7) Singla V, Galwa RP, Saxena AK, Khandelwal N (2008) Cruveilhier Baumgarten syndrome with giant paraumbilical vein. *J Postgrad Med* 54:328-9 (PMID: 18953157)
- 8) Masoodi I, Farooq O, Singh R, Ahmad N, Bhat M, Wani A (2009) Courveilhier baumgarten syndrome: a rare syndrome revisited. *Int J Health Sci (Qassim)* 3:97-9 (PMID: 21475517)
- 9) Lang H, Knitsch W, Weimann A, Manns M, Pichlmayr R (1993) Sonographic aspects of the Cruveilhier Baumgarten syndrome. *Bildgebung* 60:28-30 (PMID: 8485368)

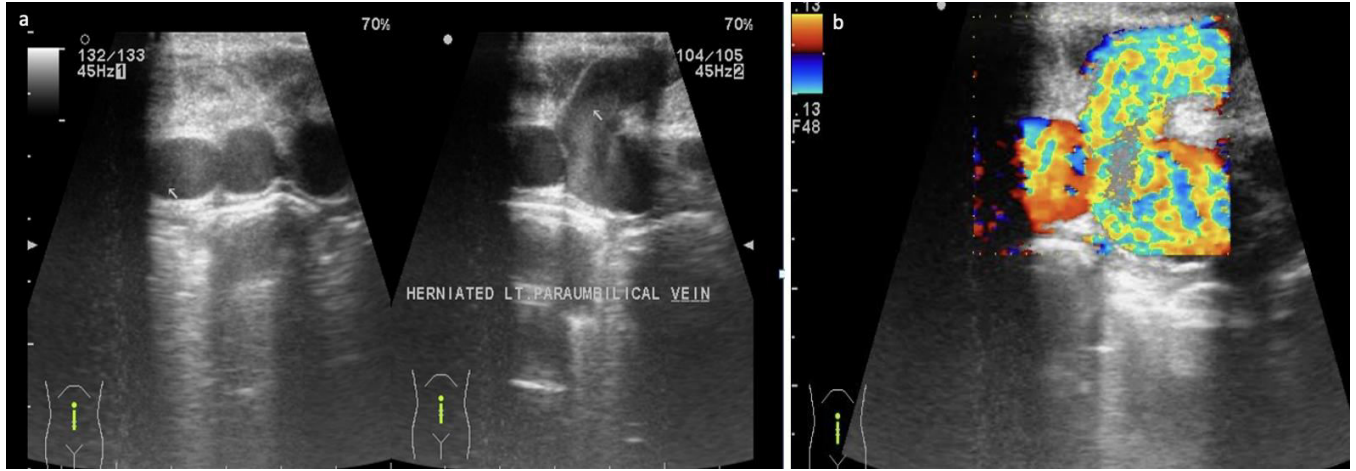


Figure 1a: Transverse 2D US sections in dual screen mode reveals herniation of left dilated paraumbilical vein through a defect in anterior abdominal wall in umbilical region

Figure 1b: Transverse US sections on color Doppler mode show the aliasing in herniated and dilated left paraumbilical vein.

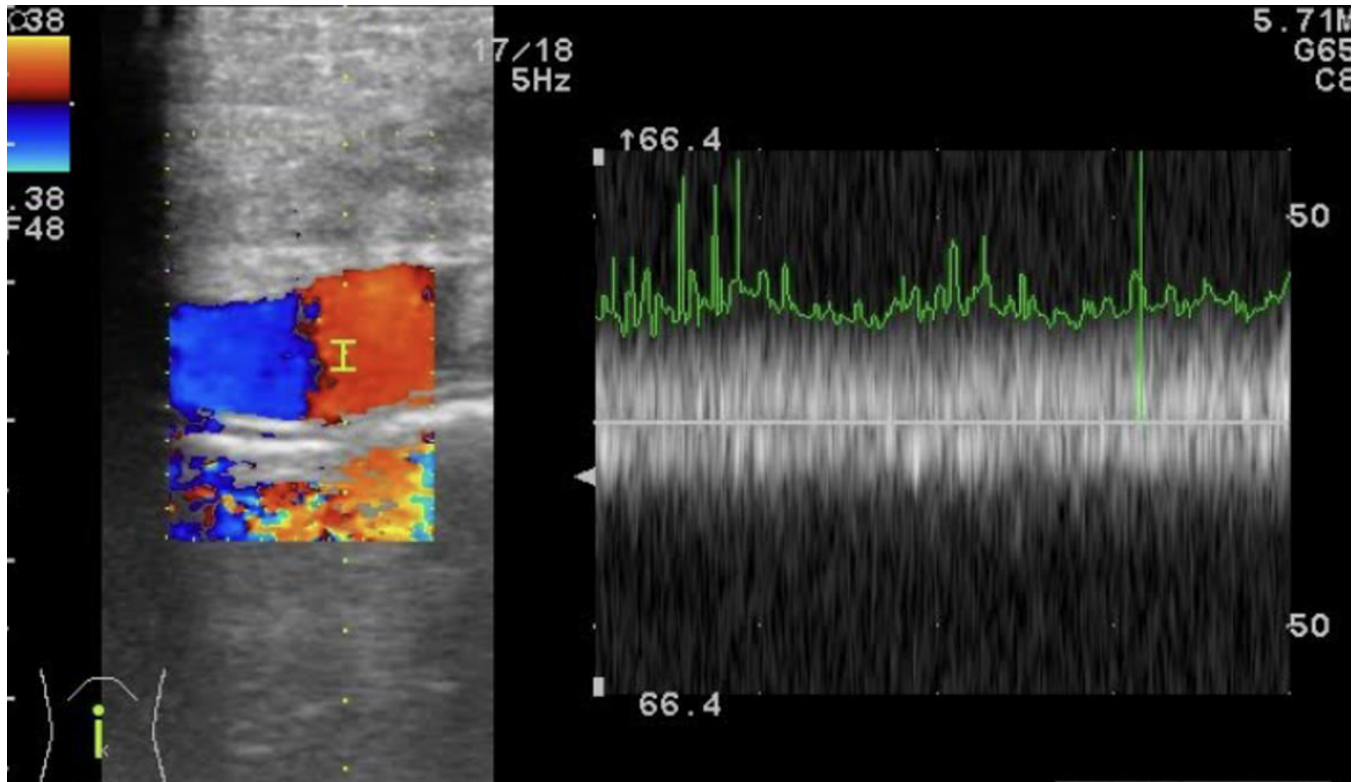


Figure 2: Transverse sections of US at the umbilical region shows dilated left paraumbilical vein. On color and spectral Doppler, it showed venous flow and waveform with a PSV of 33 cm/sec.

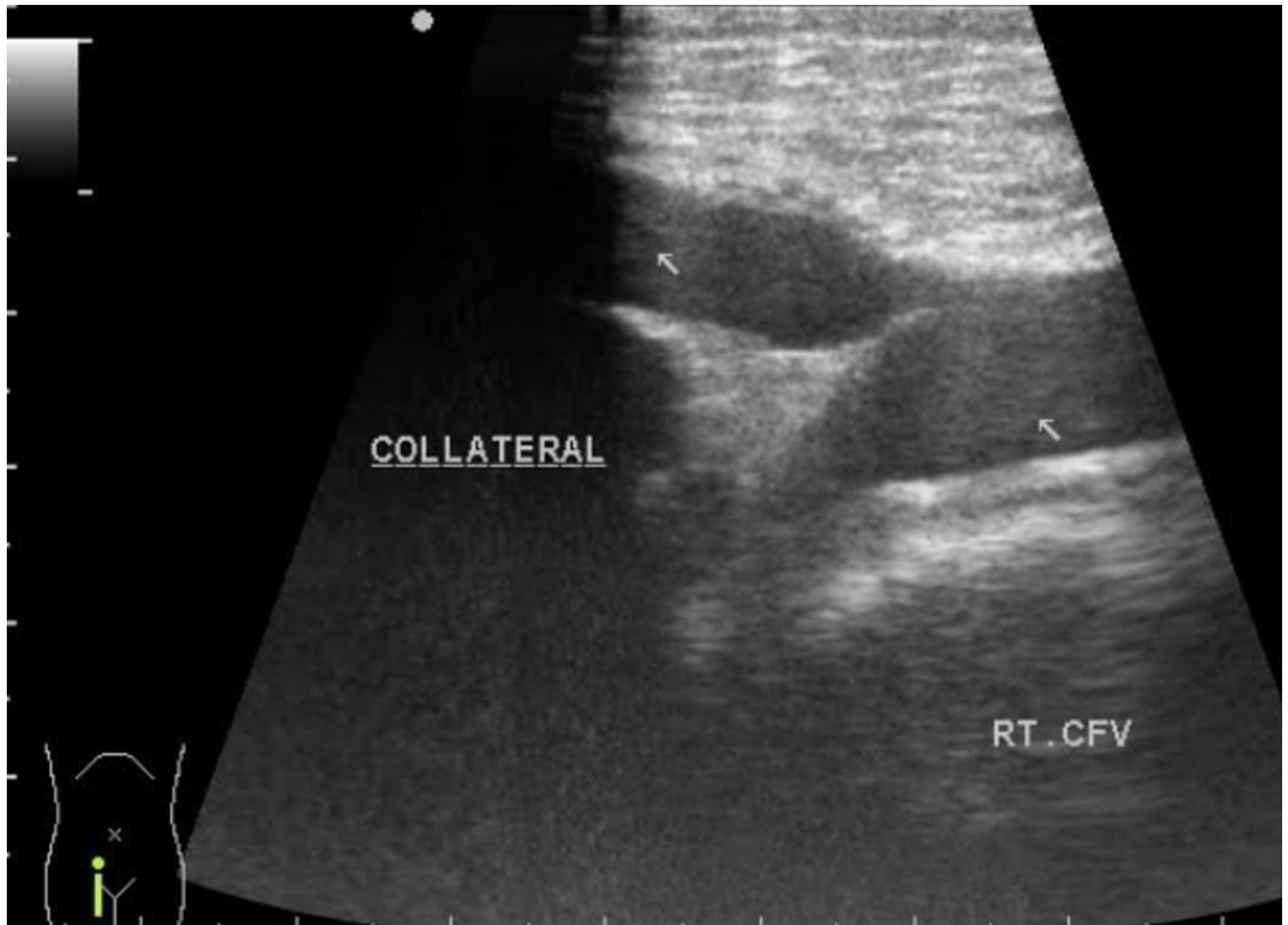


Figure 3: Transverse sections of 2D US reveal a collateral from the left paraumbilical vein draining to the right common femoral vein

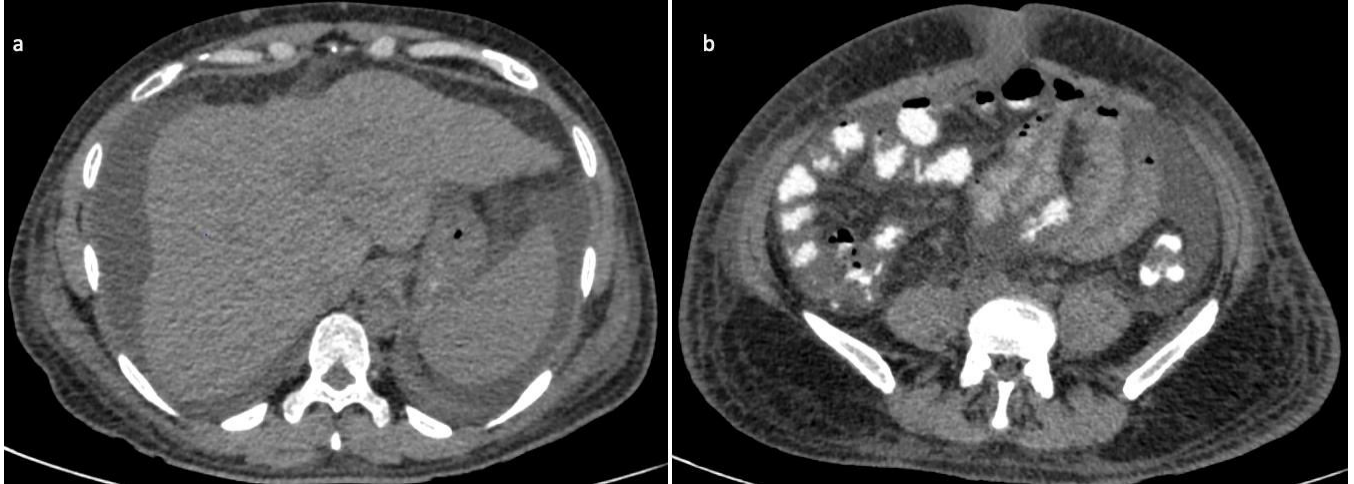


Figure 4a: Plain axial CT abdomen images reveal shrunken right lobe of liver with irregular hepatic surface and ascites.

Figure 4b: Plain axial CT abdomen images reveal ascites and herniation of paraumbilical vein through umbilicus.

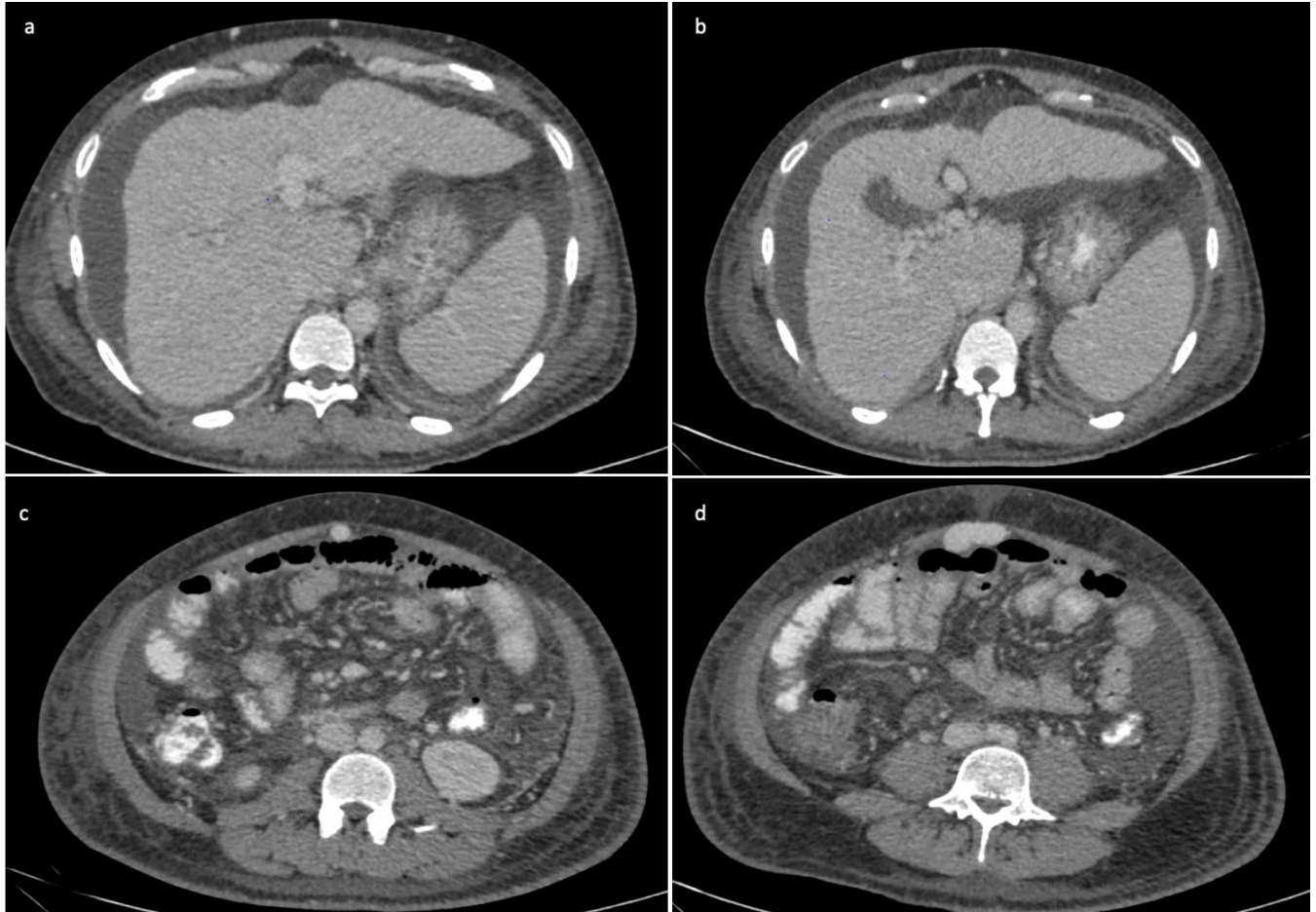


Figure 5a & Figure 5b: Axial CT abdomen post contrast images reveal shrunken right lobe of liver with surface nodularity and ascites. Multiple periportal collaterals are also noted.
Figure 5c & Figure 5d: Axial CT abdomen post contrast images reveal dilated left paraumbilical vein beneath the anterior abdominal wall in the umbilical region

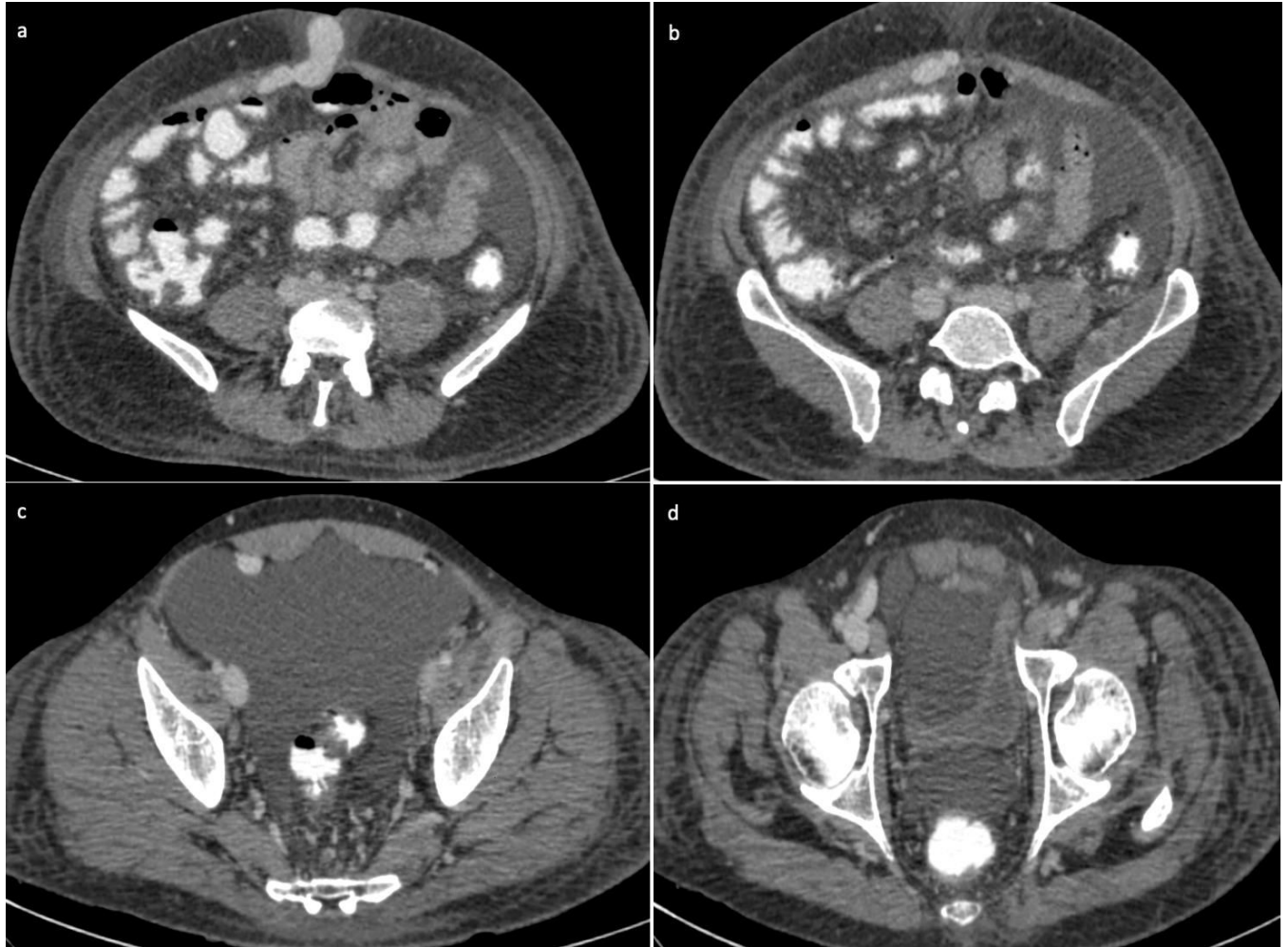


Figure 6a & Figure 6b: Axial CT abdomen post contrast images reveal herniated left paraumbilical vein through anterior abdominal wall defect in umbilical region.
Figure 6c & Figure 6d: Axial CT abdomen post contrast images reveal dilated collateral draining into the right common femoral vein.

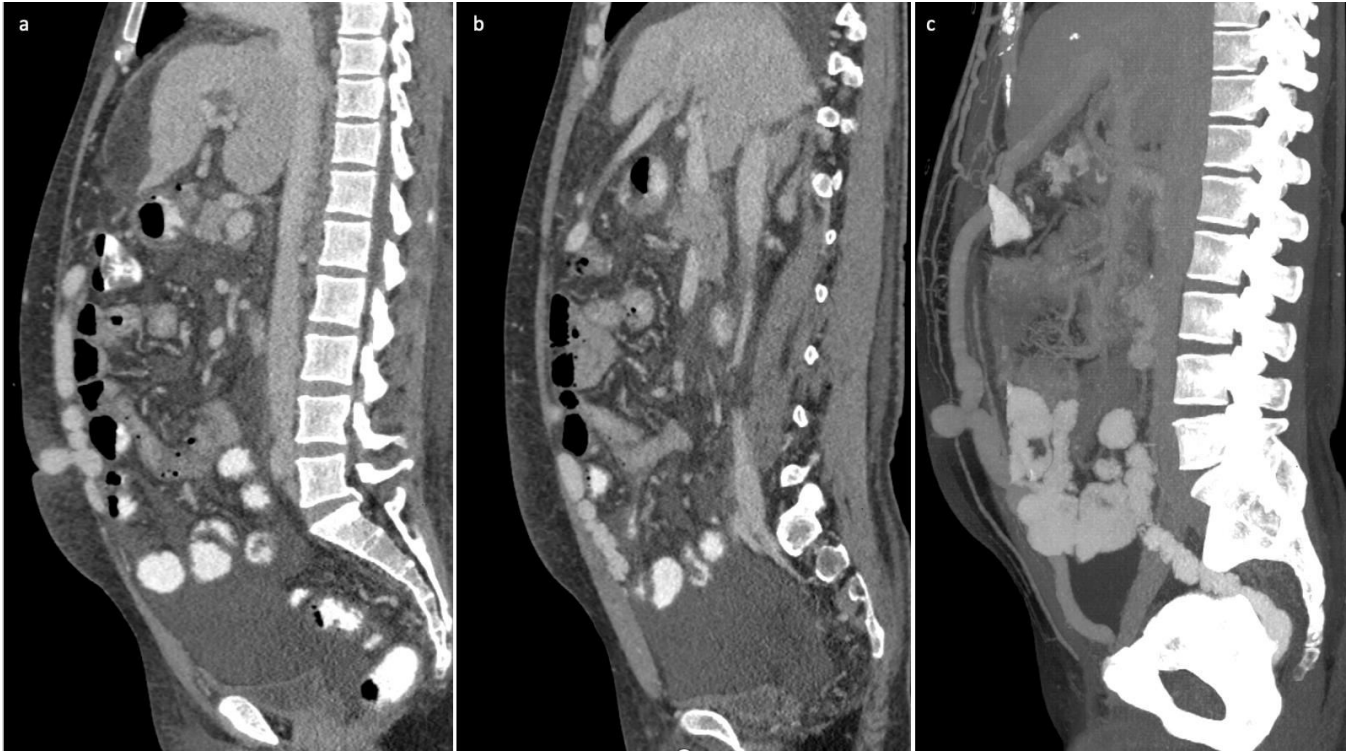


Figure 7a & Figure 7b Sagittal CT abdomen post contrast images reveal nodular liver surface, ascites and herniated left paraumbilical vein through anterior abdominal wall defect in umbilical region.

Figure 7c: Sagittal thick reformatted images of CT abdomen post contrast images reveal herniated left paraumbilical vein through anterior abdominal wall defect in umbilical region. A dilated collateral beneath anterior abdominal wall is then seen draining into right common femoral vein.

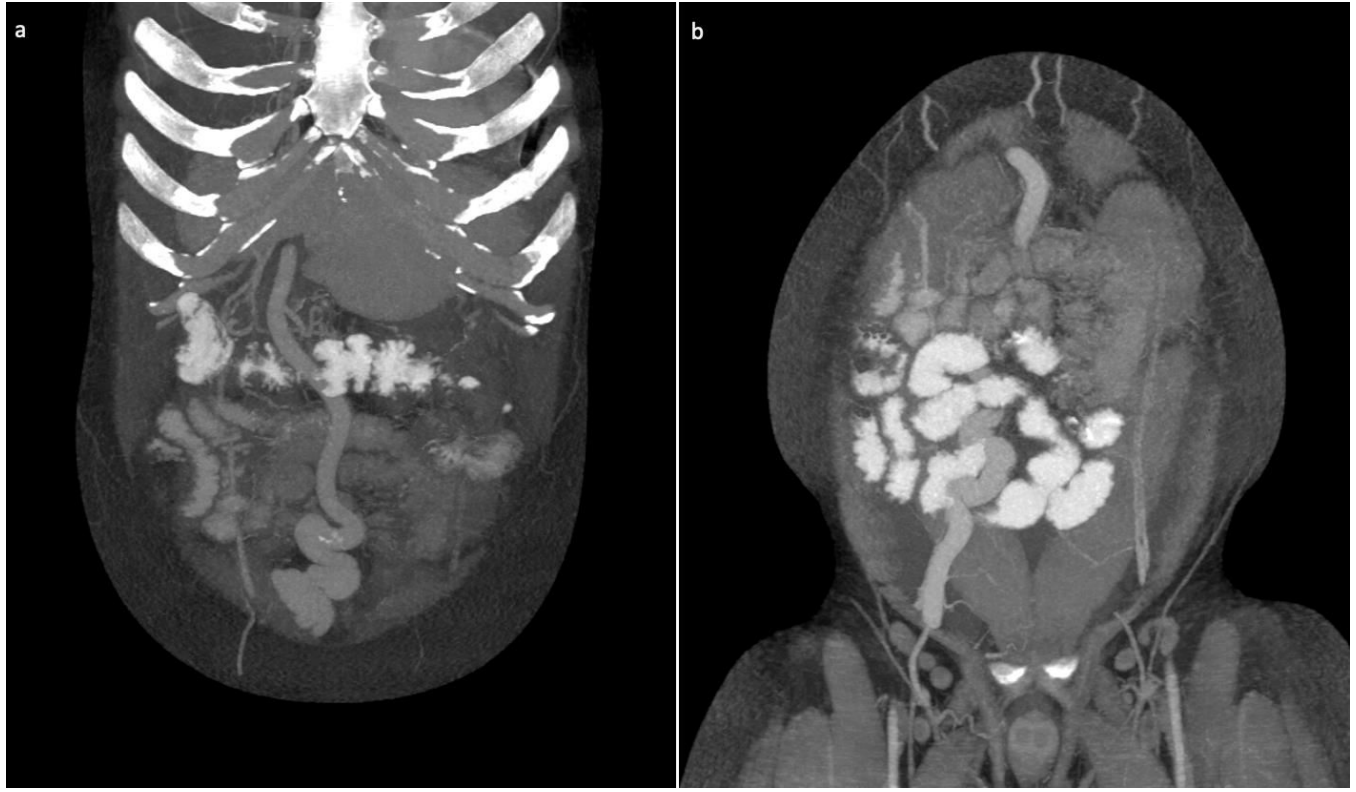


Figure 8a & Figure 8b: Coronal thick reformatted images of CT abdomen post contrast images reveal collateral from left paraumbilical vein, draining into right common femoral vein.