

Cruveilhier-Baumgarten syndrome – a caveat for surgeons

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In the presence of portal hypertension, a recanalised para-umbilical vein and splenomegaly are diagnostic of a rare condition, namely Cruveilhier-Baumgarten syndrome. In this case demonstration of the large and tortuous vein by computed tomography (CT) imaging avoided catastrophic haemorrhage from a planned hysterectomy and served as a road map for surgical planning.

Case report

A 39-year-old woman presented with lower abdominal pain and menorrhagia. Clinical examination showed pallor, hepatosplenomegaly, and a pelvic mass which was discovered to be

a multifibroid uterus. Laboratory tests were positive for bilharzia, which accounted for the hepatosplenomegaly.

In addition to the multifibroid uterus, an ultrasound showed a complex left ovarian mass, a massive splenomegaly of 20 cm, splenic varices, dilatation of the splenoportal confluence, and a large venous structure in the region of the portal vein extending inferiorly just deep to the anterior abdominal wall. It was, however, difficult to demonstrate the lower extent of this structure, so the patient underwent a CT scan.

The CT scan (Fig. 1) confirmed a dilated, serpiginous venous channel extending from the dilated left portal vein, following a course behind the anterior abdominal wall, continuing to the pelvis, taking a few turns in the suprapubic area and finally terminating in the left femoral vein. This constitutes a variant of Cruveilhier-Baumgarten syndrome. The patient did not have a significant caput medusae, and only a few small varices could be seen anastomosing with slightly distended inferior epigastric veins.



Fig. 1. Coronal three-dimensional reconstruction of the CT scan shows the dilated para-umbilical vein, its tortuous course and termination in the left femoral vein. Also note the massive splenomegaly.

Discussion

The umbilical vein becomes the ligamentum teres after birth. In portal hypertension, collaterals that develop in the ligamentum teres could recanalise it to form a porto-systemic shunt. (These collaterals would then be called a para-umbilical vein.) A caput medusae forms around the umbilicus, then draining into the systemic circulation via the inferior epigastric veins. This is only one of several portosystemic shunts that may develop in portal hypertension.²

Cruveilhier-Baumgarten syndrome is a rare syndrome characterised by portal hypertension, splenomegaly and a prominent para-umbilical circulation. It was first described in 1852 by Cruveilhier and then in 1908 by Baumgarten.^{3,4} Classic Cruveilhier-Baumgarten syndrome is characterised by a portosystemic shunt from the left portal vein via a dilated para-umbilical vein that terminates at the umbilicus in a caput medusae.³

Our patient developed portal hypertension secondary to bilharzia. The para-umbilical vein was large and tortuous, not terminating at the umbilicus but extending into the pelvis to terminate in the left femoral vein, which is an unusual variant of Cruveilhier-Baumgarten syndrome. It became clear from the

imaging that either a midline or a Pfannenstiel incision would have resulted in catastrophic haemorrhage. Identification of this abnormal vessel helped to avoid serious complications during hysterectomy and provided a surgical 'road-map'.

Conclusion

Pre-surgical imaging is an elegant way of diagnosing pathology related to presenting symptoms and signs. It is also useful for demonstrating any associated unrelated incidental pathology and for providing a 'road-map' for surgery. In our case, the demonstration of a massive para-umbilical vein was helpful

in avoiding incisions that could have resulted in catastrophic haemorrhage.

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Prevention of hepatocellular carcinoma

continued from page 51

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