Case report

Cruveilhier-Baumgarten (C-B) disease

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SUMMARY A case of Cruveilhier-Baumgarten (C-B) disease is described. It is the first reported from Zambia and from Southern Africa.

The C-B disease with a patent umbilical vein, portal hypertension, and without significant changes in the liver is a rare clinical entity. The diagnosis in the present case was made by a splenovenogram and liver biopsy. The patient was to be readmitted for ultra-sound but died in his village before this could be done.

Case report

A 47 year old Zambian man (CM), an office orderly, presented with complaints of an abdominal 'lump' of about six months duration and generalised abdominal distension for about 15 years (Fig. 1). There was no weight loss, haematemesis, or melaena, or previous history of jaundice, no blood transfusion and no alcoholic intake. He had mild pallor, but no spider naevi or palmar erythema. Grossly dilated veins in the anterior abdominal wall, more on the left side were present with a large coil of veins immediately above the umbilicus. A palpable thrill with a venous hum was heard over the varix around the umbilicus. The direction of the flow in the veins was upwards from the umbilicus. There was a para-umbilical hernia. The spleen was markedly enlarged to the level of the umbilicus. The liver was small and palpable in the epigastrium. There was ascites with gross abdominal distension.

Investigations revealed a pancytopenia with a haemoglobin 10.5 g/dl, a total white cell count $1.4 \times 10^9/l$, platelets $45 \times 10^9/l$ and slightly impaired

Fig. 1 Generalised abdominal distension with grossly dilated veins and varix around umbilicus.

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Fig. 2 Splenoportogram showing patent umbilical vein (arrowed).

coagulation (prothrombin concentration 88%). The liver function tests were slightly abnormal – total protein 88 g/l, albumin 39 g/l, bilirubin total 25.5 μ mol/l (conjugated 20.4 μ mol/l), SGOT 23 IU/l, alkaline phosphatase 213 IU/l. The VDRL and HBsAg were negative. Urine and stool tests were normal. Stools for occult blood were negative. Ascitic fluid was a transudate. Chest x-ray was normal. Oesophageal varices and mild gastritis were seen on endoscopy. The splenoportogram showed a patent umbilical vein (Fig. 2). Liver histology showed normal lobular architecture with increased inflammatory cells in the portal triads (hepatitis), prominent Kupffer cells and the presence of mononuclear cells in the sinusoids with no evidence of cirrhosis.

Discussion

Cruveilhier-Baumgarten disease is a rare condition, characterised by the presence of prominent umbilical or para-umbilical veins, abdominal venus hum with thrill, splenomegaly, normal or small liver without evidence of cirrhosis, portal hypertension with oesophageal varices and hypersplenism. Pegot (1833)¹ in Paris first reported a case of distended veins of the

abdominal wall and venus hum from around the umbilicus. Cruveilhier (1835)² and Baumgarten (1907)³ reported two cases.

Armstrong *et al*⁴ with modifications by Steinburg and Galambos,⁵ identified two different clinical entities: (1) the C-B syndrome where primary liver disease, mainly cirrhosis or portal hypertension is responsible for the extensive dilatation of the para-umbilical veins and recanalisation of the umbilical vein and (2) C-B disease where the clinical picture is the result of a congenitally patent umbilical vein with little demonstrable liver disease or portal tract abnormality.

The prominent features of the slightly abnormal liver function tests in this patient (caused by hepatitis of unknown cause) were hyperbilirubinaemia especially in the conjugated fraction and raised alkaline phosphatase activity. In view of the hepatitis the patient was treated medically and portocaval shunt for portal decompression was not indicated. He was discharged on diuretics, antacids and vitamin supplements and was to be readmitted for ultrasound at a later date.

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References

- 1 Pegot N. Tumeur variqueuse avec anomalie du système veineux et persistance de la veine ombilicale, développement des veins sous-cutanées abdominales. *Bull Soc Anat (Paris)* 1833; **8**: 57.
- 2 Cruveilhier J. *Anatomie pathologique du corps humain*. Paris: JB Baillière, 1835.
- 3 Von Baumgarten P. Uebervoll standiges Offenbleiben der Vena umbilicalis; zugleich ein Beitrag zur Frage des Morbus Bantii. *Arb Geb Pathol Anat Int Tubingen* 1908; **6**: 93–110.
- 4 Armstrong EL, Adams WL, Tragerman LJ, Townsend EW. The Cruveilhier-Baumgarten syndrome. *Ann Intern Med* 1942; **16**: 113–51.
- 5 Steinberg JS, Galambos JT. Cruveilhier-Baumgarten (C-B) Disease: an experiment of Nature. *Am J Med* 1967; **43**: 284–8.