

THE CRUVEILHIER-BAUMGARTEN SYNDROME

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INTRODUCTION

THE CRUVEILHIER-BAUMGARTEN Syndrome is a rare clinical entity characterised by the presence of unusual prominent para-umbilical veins, evidence of portal hypertension, splenomegaly and the demonstration of a venous hum, frequently with a thrill, at the site of the para-umbilical circulation. In the syndrome, the umbilical vein is patent as a result of primary liver disease. When the patency of the umbilical or para-umbilical vein is not associated with recognizable significant structural abnormalities of the liver, it is called the Cruveilhier-Baumgarten Disease.

Since the first case report (Pegot, 1933), only 86 cases of Cruveilhier-Baumgarten Syndrome have been described in the literature until 1954 (Cheng *et al.*, 1954). Since then, sporadic case reports have appeared in the English literature (Dikshit *et al.*, 1966; Banerjee, 1969; Fung *et al.*, 1971). The only previously reported case of the Cruveilhier-Baumgarten Syndrome in this region has been that of Fung, Tock and Yu (1971) in a 56 year old Chinese man from Singapore.

We report a further 2 cases of the syndrome, one in a Malay child and the other in an Indian adult, seen in Medical Unit 4, General Hospital, Kuala Lumpur in the past nine months.

CASE REPORT 1

A 14-year old Malay boy was admitted to Old Ward 13, General Hospital in March 1978 with spastic paraplegia for over 10 years. The consul-

tant neurologist felt that this was probably due to congenital absence of cortico-spinal tracts within the spinal cord. The neurological investigations including a myelogram was normal. Other relevant physical signs included congenital absence of digits of left hand; a pigeon chest deformity and a ventricular septal defect with cardiomegaly.

There was no jaundice no spider naeri or palmar erythema. There was grossly dilated veins in the anterior abdominal wall especially around the umbilicus with a large coil of veins 6 cm above the umbilicus. (Fig. 1). The direction of flow of the veins was away from the umbilicus. A palpable thrill was present over the varix above the umbilicus and on auscultation a loud continuous murmur was audible. The liver was not palpable; the spleen was enlarged to the level of the umbilicus. No ascites was detected.

Investigations revealed pancytopenia (haemoglobin, 6.8 gm/100 ml; platelets, 34,000/cu. mm total white cell count, 1,650/cu. mm). Urea and electrolytes were normal. The total bilirubin was 1.3 mg %; alkaline phosphatase, 9 KA units; total proteins, 5.3 gms. % (albumin was 3.1 gm % globulin was 2.2 gm %); SGPT was 2RF units. The prothrombin time was 2 seconds prolonged. The HBs Ag was negative. Barium swallow did not demonstrate any oesophageal varices. The splenoportogram (Fig. 2) showed dilated splenic and portal veins with a large patent umbilical vein feeding large varices in the anterior abdominal wall. The liver biopsy showed macronodular cirrhosis.

CASE REPORT 2

A 38 year old Indian gardener was first admitted into Old Ward 13, General Hospital, Kuala Lumpur in December, 1977 with the complaints of abdominal distension, yellow discolouration of eyes, swelling of both feet and loss of appetite for 2 years. No history of hematemesis or malaena. His consumption of alcohol has been

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Fig. 1. Photograph of anterior abdominal wall showing dilated tortuous veins. The arrow indicates the site of the venous thrill.



Fig. 2. Splenoportogram of the 14-year old Malay patient, showing dilated portal and splenic veins with a large patent umbilical vein feeding the periumbilical varices.

heavy for the past 25 years; his average weekly intake includes 4 pints of beer, 10 pints of toddy and 2 small bottles of chinese wine. Physical examination revealed a mildly jaundiced male with pitting ankle oedema with no spider naevi or palmar erythema. His abdomen was distended with ascites but no dilated periumbilical veins were detected then. Liver was palpable 2 cm below right costal margin; spleen was not palpable. He was diagnosed as having alcoholic liver cirrhosis with portal hypertension and was treated with diuretics and Vitamin K. He was advised to refrain from alcohol consumption.

The patient defaulted follow-up and was re-admitted on 17th March 1978 with similar complaints. Examination revealed similar findings as before except that he now had dilated periumbilical veins. Though a bruit was not palpable, a continuous venous hum was audible on auscultation just above the umbilicus.

Laboratory investigations showed a haemoglobin of 11.6 gm %; a platelet count of 74,000/cu. mm with a normal white cell count. Urea and electrolytes were normal. Liver function tests showed a serum bilirubin of 1.7 mg %, serum alkaline phosphatase of 6.0 K.A. units, serum albumin of 2.9 gm %, serum globulin of 5.3 gm %, SGPT of 2 RF units. The prothrombin time was 2 seconds prolonged. The hepatitis B surface antigen (HBsAg) was negative. Barium swallow did not demonstrate any oesophageal varices. Three days after admission the patient developed a spontaneous bleed from his umbilical varices, loosing about 250 ml of venous blood. Only when the bleeder was ligated did the bleeding stop. No blood transfusion was required.

A splenoportogram (Fig. 3) showed a slightly dilated portal vein with a patent umbilical vein feeding the varices around the umbilicus. A liver biopsy showed micronodular cirrhosis.

The patient was discharged with diuretics and vitamin supplement. He is being followed up in the hepatothology clinic.

DISCUSSION

The Cruveilhier-Baumgarten Disease is rare. Until 1972 only eleven cases of the disease have been recorded (Ozsoylu *et al.*, 1972). Unlike the Cruveilhier-Baumgarten Syndrome, the liver is



Fig. 3. Splenoportogram of the 38-year old Indian patient, showing the patent umbilical vein (arrowed) feeding the umbilical varices.

morphologically normal with no cirrhosis. Our 2 cases are those of the Cruveilhier-Baumgarten Syndrome as they both had underlying liver disease (cirrhosis) to account for the prominent para-umbilical veins with the demonstrable venous hum. In the clinical diagnosis of the disease or the syndrome, the venous hum is of great importance. The mechanism of the production of the murmur is still unsettled, although its dependence upon the venous anastomoses is beyond doubt.

In both our patients the venous hum was detected and a patent umbilical vein feeding the umbilical varices was demonstrated in each case with splenoportogram. Splenomegaly has been detected in most of the previously reported cases. The spleen was enlarged in our Malay boy but was not palpable in our second case. Anaemia, leukopenia and thrombocytopenia (hypersplenism) was evident in our patient with the clinically enlarged

spleen but not in our Indian patient. Eosinophilia was absent in both our cases. Splenomegaly with or without eosinophilia, hepatomegaly, anaemia and persistent leukopenia have been stated as additional clinical criteria for the diagnosis of Cruveilhier-Baumgarten Syndrome. (Cheng *et al.*, 1954). Our first case is interesting because we believe that this is the first case of a young Malay boy with the Cruveilhier-Baumgarten Syndrome associated with multiple congenital deformities (pigeon-chest, absent digits, ventricular septal defect and perhaps congenital abnormalities in the spinal cord).

We feel that routine clinical auscultation of the periumbilical region in all patients with portal hypertension is necessary to pick up all cases of the Cruveilhier-Baumgarten Syndrome.

SUMMARY

Two cases of the relatively uncommon Cruveilhier-Baumgarten Syndrome are reported; one in a 14-year old Malay boy; the other in a 38-year old Indian man, both with cirrhosis of the liver. The 14-year old boy had associated multiple congenital abnormalities. Some important clinical features of the Syndrome are discussed. The need for routine auscultation of periumbilical region in patients with portal hypertension is emphasised.

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