

CASE REPORT

CRUVEILHIER-BAUMGARTEN SYNDROME IN A CIRRHOTIC PATIENT TAKING ALCOHOL WITH *BOESENBERGIA PUNDURATA*

Polrat Wilairatana¹, Noppadon Tangpukdee¹ and Lertchai Jittaseree²

¹Department of Clinical Tropical Medicine, Faculty of Tropical Medicine, Mahidol University, Bangkok; ²Bang Pa-in District Hospital, Ayutthaya, Thailand

Abstract. Cruveilhier-Baumgarten syndrome (CBS) is a rare complication of cirrhosis. We reported a case of CBS who came to hospital with progressive ascites, jaundice, fever and upper gastrointestinal bleeding. The cause of cirrhosis and hepatitis might be due to alcohol and possibly local Thai herb [*Boesenbergia pundurata* (Roxb.) Schitr or Krachaidum].

Key words: Cruveilhier-Baumgarten syndrome, cirrhosis, alcohol, *Boesenbergia pundurata*

INTRODUCTION

Cruveilhier-Baumgarten syndrome (CBS) is a rare medical condition of liver cirrhosis or portal hypertension with distension of the paraumbilical vein (*ie*, an acquired condition in which the veins reopen due to high portal pressure). The Cruveilhier-Baumgarten disease or Pégot-Cruveilhier-Baumgarten disease is a term reserved for cases with distension of the paraumbilical veins due to failure of umbilical vein closure, with little or no evidence of liver disease found on liver biopsy (*ie*, a congenital patency of the umbilical vein leading to portal hypertension).

We reported a rare case of CBS in Thai-

land in a cirrhotic patient. The cause of cirrhosis and hepatitis might be alcohol and possibly a local Thai herb. Subsequent spontaneous bacterial peritonitis (SBP) contributed to liver function deterioration and might also cause jaundice.

CASE REPORT

A 32 year old Thai man was admitted to the hospital with a history of progressive abdominal distension, jaundice, fever and hematemesis with melena for 1 month, 2 weeks, 3 days, and 1 day prior to hospital admission, respectively. The patient noticed progressive vein dilatation on the abdominal wall above the umbilicus for 1 year. He had taken alcohol with a local Thai herb [*Boesenbergia pundurata* (Roxb.) Schitr or Krachaidum], 30-60 ml (1 *Pek* in Thai) per day for 3 years and had no history of altered sensorium.

On examination, the patient was conscious, oriented and had an oral tempera-

Correspondence: Dr Polrat Wilairatana, Department of Clinical Tropical Medicine, Faculty of Tropical Medicine, Mahidol University, 420/6 Ratchawithi Road, Ratchathewi, Bangkok 10400, Thailand.

E-mail: tmpwl@mahidol.ac.th



Fig 1–The patient had jaundice and marked icteric sclera.

ture of 37.8°C. He also had pallor with icteric sclera (Fig 1), jaundice, spider nevi over the left chest wall, tattoos on the right shoulder, left chest wall, and left arm (Fig 2). Gynecomastia was observed (Figs 2 and 3). The abdomen showed a liver span of 11 cm, no palpable spleen, ascites and positive bowel sounds without rebound tenderness. Grossly dilated tortuous veins above the umbilicus with a venus hum and thrill were detected (Fig 2). Pitting edema was observed of the pretibial area of both legs. A complete blood count showed a hemoglobin of 10.5 g/dl (normal value 13.3-16.2 g/dl), a white blood count of $4.6 \times 10^3/\text{mm}^3$ (comprised of 54% neutrophils, 4% lymphocytes, 1% monocytes and 1% eosinophils), and a platelet count of $48 \times 10^3/\text{mm}^3$. Liver function test showed:



Fig 2–Generalized abdominal distension due to ascites with grossly dilated veins above the umbilicus. Ecchymoses were seen at the venepuncture sites of both anticubital fossae and at abdominal paracentesis site at the right lower abdomen. Gynecomastia and tattoos (of the right shoulder, left chest wall, and left arm) are also seen.



Fig 3–Spider nevi of the left chest wall, gynecomastia and tattoos are observed.

total bilirubin/direct bilirubin of 40.3/24.1 mg/dl (normal values 0.1-1/0-0.3 mg/dl), AST/ALT of 355/91 U/l (normal values <37/<41 U/l), alkaline phosphatase of 137 U/l (normal value 57-128 U/l), albumin/globulin of 2.8/4.2 mg/dl (normal values 3.5-5/1.8-3.4 mg/dl) and BUN/Cr of 5/1.7 mg/dl (normal values 6-20/0.6-1.3 mg/dl). Nasogastric aspiration showed 7.5 ml of coffee ground fluid but no fresh blood detected. Coagulation study, upper gastroendoscopy, abdominal ultrasonography, hepatotropic virus study, phonovenogram or stethogram of the abdominal venous hum of the large tortuous paraumbilical veins, bacterial culture of the blood and ascitic fluid were not attempted due to limited facilities at the district hospital. An abdominal paracentesis was conducted, but the results were not recorded. The initial diagnosis in this patient was CBS, cirrhosis with ascites, SBP, and upper gastrointestinal bleeding.

The patient was treated with ceftriaxone 2 g iv once daily combined with metronidazole 500 mg iv 8 hourly for SBP, omeprazole 40 mg iv 12 hourly for upper gastrointestinal bleeding, and spironolactone 25 mg twice daily for reducing ascites. After antibiotic treatment, the fever gradually subsided. On day 5 of admission the patient became afebrile and the stool became yellow. However, the patient still had abdominal discomfort due to ascites, which was not decreased much with the diuretic, and he needed a further abdominal paracentesis to relieve his abdominal discomfort.

DISCUSSION

Armstrong *et al* (1942) found the patients with CBS had primary liver disease, mainly with cirrhosis or portal hypertension, which was responsible for the exten-

sive dilatation of the paraumbilical veins and recanalization of the umbilical veins. The patient in this report had clinical cirrhosis with spider nevi, gynecomastia, ascites, a reversed serum albumin/globulin ratio, and possibly hypersplenism contributing to thrombocytopenia. Anemia might be due to upper gastrointestinal bleeding and possibly hypersplenism. The patient had a bleeding tendency (from thrombocytopenia and possibly coagulopathy) contributing to ecchymoses at abdominal paracentesis site and the venepuncture sites at the antecubital fossae (Fig 2). Pretibial pitting edema was due to hypoalbuminemia. The cause of upper gastrointestinal bleeding was unknown since upper gastroendoscopy was not attempted, however bleeding was not noted by nasogastric fluid aspiration on admission. Liver function test in the patient showed hyperbilirubinemia with elevation of both AST and ALT.

The finding of AST>ALT might be due to alcoholic hepatitis or toxic hepatitis, possibly from the herb the patient had taken in a large amount daily for 3 years. Some herbs are toxic for some people and not toxic for others. Some Thai people take *Boesenbergia pumdata* (Roxb.) Schitr or *krachaidum*, since they believe this herb can make them healthy and strong, similar to ginseng, help ejecting air from stomach, help with digestion, relieve a bloated stomach, gassy stomach and queasiness, nourish the heart, eject urine and leukorrhea (in females). Although there have been no reports of *krachaidum* toxicity in the literature. It was possible this herb was hepatotoxic to this patient since the patient's liver was abnormal due to chronic alcoholism. Although the patient took daily alcohol for only 3 years, consumption in large amounts might cause alcoholic hepatitis and cirrhosis. Another

possible cause of jaundice in this patient was infection due to SBP which could worsen the liver function. Although laboratory diagnosis of SBP was not obtained, the fever gradually subsided after parenteral antibiotic treatment for 5 days, which suggested the possible diagnosis of SBP. Hepatitis from hepatotropic viruses (hepatitis A, B, and C) is unlikely since ALT in viral hepatitis is usually greater than AST. The elevated alkaline phosphatase might be due to cholestasis rather than surgical obstructive jaundice. Ultrasonography of the biliary system to exclude biliary tract obstruction was not conducted in this patient due to investigation constraints of the hospital. Cirrhosis in this patient could not be clinically staged using the modified Child-Pugh classification since a prothrombin time was not obtained (the Child-Pugh score is calculated from 5 factors: serum bilirubin, serum albumin, prothrombin time, ascites and hepatic encephalopathy).

In classic CBS, the umbilical portion of left portal vein feeds a paraumbilical vein, which leaves the liver and then heads towards the umbilicus (Morin *et al*, 1992; Singla *et al*, 2008). The paraumbilical vein is seen as a tubular structure arising in the falciform ligament between the left lobe of liver, leading from the left portal vein to veins of the anterior abdominal wall (Palazon and Minguez, 1997). The development of a large recanalized paraumbilical vein has been found to prevent formation of bleeding esophageal varices and predisposes to hepatic encephalopathy (Lam *et al*, 1981). These subcutaneous collaterals may undergo spontaneous hemorrhage or inadvertent significant bleeding during abdominal paracentesis

(Sodhi *et al*, 2007; Singla *et al*, 2008).

CBS is a rare complication of cirrhosis. In Thailand there is only one previous report in the English literature (Wilairatana and Wilairatana, 1993). The present report is the second report in Thailand.

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