CASE REPORT

HEPATOPORTAL SCLEROSIS WITH ISCHEMIC BOWEL DISEASE: A RARE CASE DIAGNOSED BY CLINICAL AUTOPSY

Bommanahalli BP[1], Vidyavathi K[2], Harendrakumar ML[2]
[1] Department of Pathology, Navodaya Medical College, Raichur, Karnataka, India
[2] Department of Pathology, Sri Devaraj Urs Medical College, Kolar, Karnataka, India

Correspondence to: Bommanahalli BP (basupath@rediffmail.com)

DOI: 10.5455/ijmsph.2014.160220142 **Received Date: 04.02.2014** **Accepted Date: 16.03.2014**

ABSTRACT
A 35 year old female presented with pain abdomen, massive hematemesis and signs of portal hypertension. Death occurred despite of all efforts to manage shock & hematemesis by endoscopic sclerotherapy. Clinical autopsy revealed gangrene of small intestine with mesenteric vein thrombosis; oesophageal varices; hepatoportal sclerosis (HPS) of liver with regenerative nodules and chronic venous congestion of spleen. Histopathology of liver is essential for diagnosis of HPS & to rule out other disease process. Gangrene intestine in HPS is a rare presentation.
Key Words: Hepatoportal Sclerosis (HPS); Portal Hypertension; Gangrene; Intestine

Introduction

Non- cirrhotic portal hypertension (NCPH) is poorly understood condition, characterized by presence of portal hypertension in the absence of cirrhosis. Guido Banti, described it as morbus Banti in the late nineteenth century with a pattern of anemia, gastrointestinal bleeding and marked splenomegaly.[1] Over the years, Mikkelsen et al[2] called it as hepatoportal sclerosis. Various synonyms are used to refer this entity: idiopathic portal hypertension in Japan, non- cirrhotic portal fibrosis in India, regenerative nodular hyperplasia, benign intrahepatic portal hypertension etc.[3] We present a case of hepatoportal sclerosis with ischemic bowel disease, diagnosed by clinical autopsy.

Case Report

A 35 year old female from lower socioeconomic status, presented with history of massive hematemesis and pain abdomen since 3 hours. Physical examination revealed signs of portal hypertension (mild to moderate ascites, splenomegaly and prominent periumbilical veins) and severe pallor. Abdomen was tender with guarding rigidity. Patient was non-alcoholic and the drug history was not relevant. She was on vasoactive drugs for similar episodes of mild hematemesis.

Haematological investigations revealed 4 gm/dl of haemoglobin, low normal total leukocyte count (4200 cells/cumm) and microcytic hypochromic blood picture. Bleeding time, Prothrombin time (PT) and Activated Partial Thrombin Time (APTT) were within normal limits. Biochemical investigations showed normal liver function tests except mild rise in Aspartate aminotransferase (120 IU/L). Serological tests for Hepatitis B, Hepatitis C, HIV and ANA (antinuclear antibodies) were negative. Patient died 4 hours after admission despite of all efforts to treat hematemesis (by endoscopic sclerotherapy) and shock. Clinical autopsy was performed to know the pathology of disease after obtaining formal consent.

Autopsy Findings

Body was opened by 'I' incision from chin to symphysis pubis. Exposure of peritoneal cavity showed 750 ml of haemorrhagic fluid and gangrenous bowel. All organs were removed en mass and dissected separately.

Gross findings: Dilated tortuous veins were seen at gastro-oesophageal junction extending 6 cm proximally and, along lesser and greater curvature of stomach. On cut opening the stomach, 800 ml of coffee brown coloured fluid was found. Duodenum was unremarkable. A segment (2 meter length) of small bowel was gangrenous with reddish brown serosal and mucosal surface. The intestinal contents were reddish brown. Mesenteric vessels were dilated and thrombosed. Examination of rectum revealed internal haemorrhoids. Spleen (630 gm weight, 15 x 10x 5 cm) was enlarged with reddish cut surface. Splenic vein, portal vein and mesenteric veins were dilated. The diameter of portal vein was 2.5cm. Liver weighed 1,050 grams and measured 23 x 10 x 10 cm. External surface was nodular (Figure 1a). Cut surface revealed thick capsule and nodules of varying sizes from 0.5 to 1.0cm, distributed predominantly in the periphery (Figure 1b). Occasional dilated and thrombosed small to medium sized intrahepatic veins were seen. Other abdominal organs and thoracic organs were unremarkable.

Microscopical Features: Oesophagus and cardiac portion of stomach showed submucosal thrombosed veins. Sections from small intestine revealed ischemic mucosal infarction, oedematous submucosa and thrombosed mesenteric veins (Figure 2 a & b). Histopathology of spleen showed features of chronic venous congestion with thick capsule, dilated and congested sinusoids, and reduced white pulp. Sections of liver stained with haematoxylin and eosin, Van Gieson and Gomori's reticulin argentation revealed increased portal collagenous tissue, sclerosis and thrombosis of small branches of portal veins, arterialisation of portal vein and regenerative hyperplastic nodules (Figure 3 a & b). Microscopy of other abdominal organs and thoracic organs was unremarkable. Bone marrow aspirate and biopsy showed diminished iron stores and micronormoblastic erythropoiesis. Diagnosis of hepatoportal sclerosis with small bowel gangrene was derived by analysing clinical and pathological features.

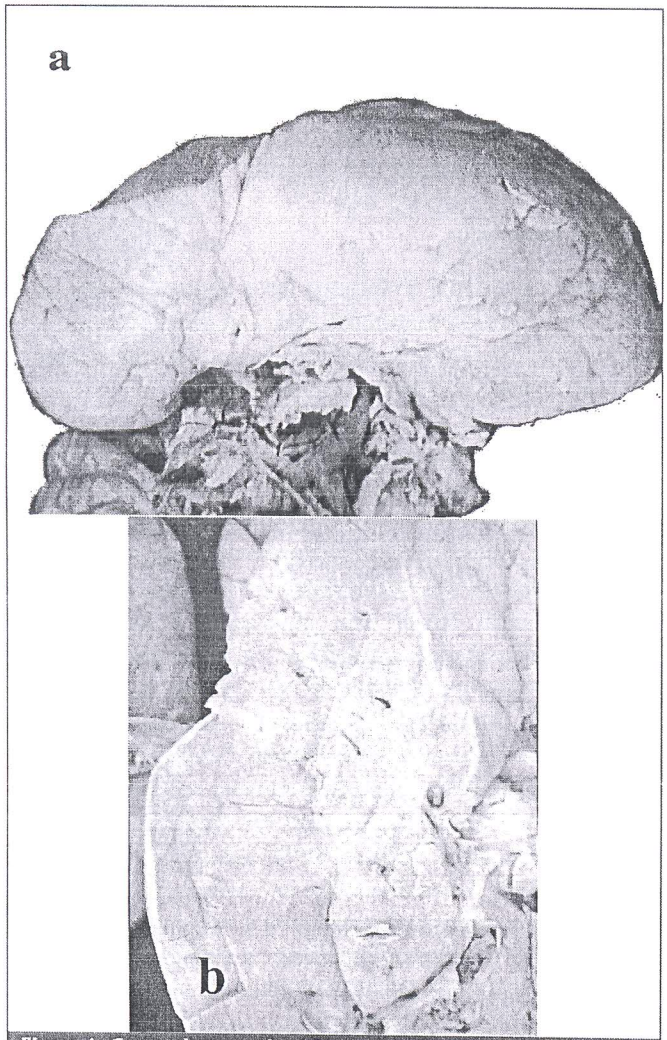

Figure-1: Gross photograph of Liver. (a) The outer surface shows nodules of varying sizes; (b) Cut surface showing nodules and branch of dilated portal vein

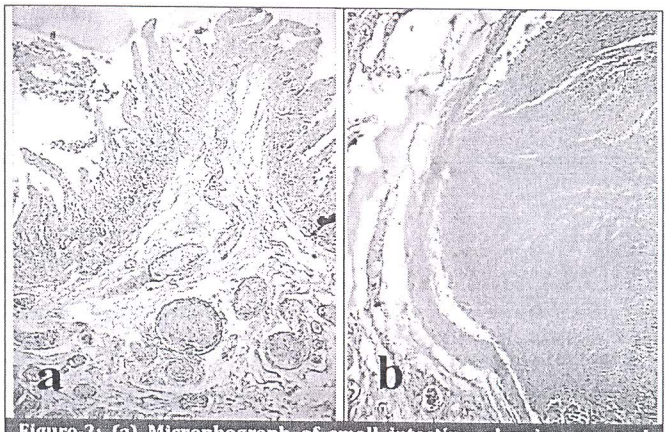

Figure-2: (a) Microphograph of small intestine: showing mucosal infarction, edematous submucosa with congested vessels; (b) Microphotograph of mesenteric vessel shows thrombus occluding entire lumen

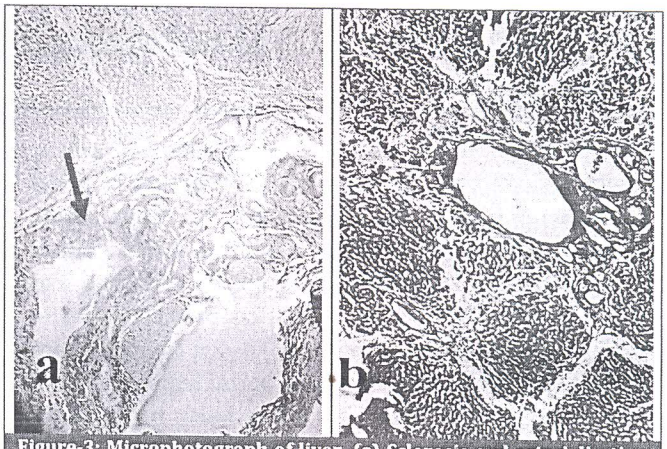

Figure-3: Microphotograph of liver. (a) Sclerosis and arterialisation of portal vein (blue arrow) (b) Regenerative nodules of varying size surrounded by reticulin fibres (Reticulin Silver nitrate, x 400)

Discussion

Hepatoportal sclerosis is reported from all over the world but the condition is more common in India and Japan. HPS is common in people of low socioeconomic status.[4,5] The peak age of occurrence is third or fourth decade of life. Studies reported male predominance or no sex predilection in India in contrary to female dominance in Japan.[6] Our case was 35 year old female belonging to low socioeconomic status.

Numbers of hypothesis were proposed to explain etiopathogenesis, although none has proven to be the cause. The factors like umbilical/ portal pyemia; diarrhoeal diseases; bacterial infections in infancy; autoimmune disorders; prothrombotic states; chronic exposure to arsenic, vinyl chloride; prolonged treatment with methotrexate; celiac disease were reported to be associated with HPS. However, exact etiology in most cases remained unknown.[7]

Clinically, most of these patients present with episodes of gastrointestinal haemorrhage, splenomegaly, anaemia, congestive gastropathy. The liver function tests may be normal or near normal with mild rise in transminases. Development of ascites, jaundice and hepatic encephalopathy, gangrene intestine is uncommon.[7] The role of prothrombotic disorders in hepatoportal sclerosis was emphasised by observations of Hillaire et al[8] that 13 of 28 patients developed extrahepatic portal vein thrombosis during follow up. Nine of these 13 cases had a prothrombotic state like myeloproliferative disorder, Polycythemia vera, protein C and protein S deficiency.[8] In our study, investigation of prothrombotic state was incomplete. We could rule out myeloproliferative disorder as prothrombotic factor by studying the bone marrow.

Histopathology of liver is essential to rule out other disease processes. Stellate or arachnoid periportal fibrosis with numerous dilated vascular channels is seen without significant intrasinusoidal collagen. Intimal fibrosis and elastosis of medium and large sized portal vein branches may obliterate the lumen and thickened veins may look like artery. Portal or parenchymal inflammatory changes are absent or minimal with normal bile ducts. Hilar and intrahepatic branches of portal vein may show eccentric luminal thickening, usually secondary to thrombosis with recanalisation. Nodular regenerative hyperplasia, focal nodular hyperplasia and incomplete septal fibrosis are also described in cases of HPS.[3,7-9]

The histopathological differential diagnoses are hereditary hemorrhagic telangiectasis, Schistosomiasis, alcoholic liver disease and fibrosis due to other causes. Telangiectatic portal lesions and honey combed meshwork of sinusoids are present in hereditary hemorrhagic telangiectasis. Classic 'pipe stem' fibrous lesion in portal vein radicals with eventual granulomatous inflammation are observed in Schistosomiasis, contrasting their absence in HPS. Presence of perisinusoidal and perivenular fibrosis, Mallory hyaline bodies, neutrophilic infiltration in lobules, differentiates alcoholic liver disease from HPS.[9]

The survival rate of patients with HPS is better than that for patients with cirrhosis. Portal vein thrombosis and ascites may indicate deterioration of the condition. Major cause of death is variceal bleeding[7] as in our case.

Conclusion

In conclusion, HPS can occur with advanced complications like massive variceal bleeding, thrombotic events (like gangrene intestine) and ascites. Study of liver histopathology & clinical correlation clinches the diagnosis of HPS, whose etiology is still unclear.

References

1. Banti BC. Ueber Morbus Banti. Folia Haematol 1910;10:33- 74.
2. Mikkelsen WP, Edmondson HA, Peters RL, Redeker AG, Reynolds TB. Extra- and intrahepatic portal hypertension without cirrhosis (Hepatoportal sclerosis). Ann Surg 1965;162:602-20.
3. Cuadrado A, Aresti S, Delgado A, Fiqols FJ, Salcines JR. Hepatoportal sclerosis: a cause of portal hypertension to bear in mind. Gastroenterol Hepatol 2008;31:104-5.
4. Dhiman RK, Chawla Y, Vaisishta RK, Kakkar N, Dilawari JB, Trehan MS, et al. Non-cirrhotic portal fibrosis (Idiopathic portal hypertension): experience with 151 patients and a review of the literature. J Gastroenterol Hepatol 2002;17:6-16.
5. Pande C, Kumar A, Sarin SK. Non-cirrhotic portal fibrosis: a clinical profile of 366 patients (abstract). Am J Gastroenterol 2006;101:S191.
6. Okudaira M, Ohbu M, Okuda K. Idiopathic portal hypertension and its pathology. Semin Liver Dis 2002;22:59-72.
7. Sarin SK, Kumar A, Chawla Y, Baijal SS, Dhiman RK, Jafri W, et al. Non-cirrhotic portal fibrosis/ idiopathic portal hypertension: APASL recommendations for diagnosis and treatment. Hep Intl 2007;1:398-413.
8. Hillaire S, Bonte E, Denninger MH, Casadevall N, Cadranel JF, Lebrec D, et al. Idiopathic non-cirrhotic intrahepatic portal hypertension in the west: a re- evaluation in 28 patients. Gut 2002;51:275- 80.
9. Kanel GC, Korula J. Non-cirrhotic portal fibrosis. In: Atlas of liver pathology. 2nd edition, Elsevier Saunders: Philadelphia. 2005. p. 132- 4.

Cite this article as: Bommanahalli BP, Vidyavathi K, Harendrakumar ML. Hepatoportal sclerosis with ischemic bowel disease: A rare case diagnosed by clinical autopsy. Int J Med Sci Public Health 2014;3:522-524.
Source of Support: Nil
Conflict of interest: None declared