BUDD-CHIARI SYNDROME – A CASE REPORT

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INTRODUCTION

Budd in 1845 and later Chiari in 1899 had reported a clinical syndrome due to occlusion of hepatic vein at junction of inferior vena cava. This syndrome however was later further reported by many others. It was also later pointed out that a similar clinical picture could also be due to inferior vena cava obstruction without hepatic vein involvement. A few aetiological factors such as intraabdominal malignancies, sepsis and polycythemia were found responsible for the thrombosis of the hepatic vein and / or inferior vena cava.

This report describes a typical case of the syndrome which was later confirmed by surgery to have inferior vena cava obstruction.

CASE REPORT

Mr. K is a 37 year old Indian dental clerk who was admitted to the University Hospital after having been found unconscious in bed by his mother. Earlier, the patient had felt drowsy and went to bed early. He could not be awakened by his mother the next day. Three days before admission, he had had watery diarrhoea.

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His illness dated back 18 years before when he noticed sudden swelling of his abdomen. He was admitted to a hospital for treatment and was later discharged from hospital with diuretic.

No definite diagnosis was given. Since then he had seen his doctor on and off for medicine to keep his abdominal swelling down. Four months before this present admission, he noticed that his abdomen was becoming prominent. His appetite became poor and he had bouts of vomiting whenever he took food. There was no history of jaundice and he was not breathless. He does not drink alcohol. There was no past history of significance except for malaria when he was a child. His father died of cirrhosis of the liver and was a heavy drinker. There was no other family history of significance.

Clinical examination revealed a rather sick looking man with much generalised wasting. He was afebrile and there was no obvious jaundice, palmar erythema, flab or telangectasia. Blood pressure was 110/70 mm Hg. There was no clinical evidence of cardiac failure or pericardial effusion and the lungs were clear. Abdominal examination showed some rather interesting gross features. There was definitely gross ascites with distended abdominal veins running longitudinally across the abdominal wall. The veins were flowing upwards and the liver was found to be enlarged to 10 cm below the costal margin. It was firm but not tender. The spleen was

enlarged to 5 cm below the costal margin. There was no scrotal swelling but varicose veins were prominent.

Investigations revealed the following : Haemoglobin concentration was 9.9 gm %, pack cell volume was 31 %, mean corpuscular haemoglobin concentration was 31.9 %, total white blood cell count was 6300/cu mm with 76 % polymorphs, 15 % lymphocytes, 6 % eosinophils and 3% monocytes and the erythrocyte sedimentation rate was 97 mm in one hour. His blood sugar estimation was 128 mg %, blood urea was 66 mg %, and electrolytes were : sodium 140 m eq/l, potassium 3.2 m eq/l and chloride 108 m eq/l. Test for syphillis was negative. Liver function tests were done and were found to be normal except for serum alkaline phosphatase which was 460 i.u. Serum protein was 7.5 gm % with albumin 3 gm % and globulin 4.5 gm %. Urine examination did not reveal any abnormality. Stool examination revealed the presence of occult blood but no ova or cyst was seen. Electrocardiogram was normal and did not show any evidence of pericarditis. His chest X-ray was normal except for the raised diaphragm due to the ascites and enlarged liver. Abdominal x-ray did not show any calcification. Ba swallow and follow through was also done and this did not show any varices and the stomach was normal. Fluoroscopy was done to rule out constrictive pericarditis but this showed good cardiac pulsation. Peritoneal fluid contained 1 gm % of protein but there was no pus cell and no growth was obtained on culture. Patient had a laporatomy done and this confirmed the diagnosis of chronic Budd - Chiari syndrome due to obstruction of the intrahepatic portion of the inferior vena cava.

DISCUSSION

The obstruction of hepatic veins causing Budd - Chairi syndrome has been well documented. More recently a similar picture of Budd - Chiari syndrome has been reported to be due purely to inferior vena cava obstruction.[1]

The cause of such obstruction can be difficult to determine. Various causes have been identified in the past. This includes developmental anomaly, invasive neoplasms from surrounding organs, thrombo-phlebitis migran, portal pyaemia or clotting diseases such as polycythemia.

The syndrome usually has an acute onset and may later become chronic. Ascites, enlarged liver and prominent abdominal veins not responding to treatment are usually prominent features of this syndrome. Distention of the hepatic capsule may be responsible for the vomiting frequently experienced by patients with Budd - Chiari syndrome. Severe watery diarrhoea is a bad sign and may indicate a terminal stage of the illness. This is said to be due to mesenteric vein obstruction. As a whole, patients tend to survive very long without treatment in spite of the gross features of the disease. Our patient has so far survived 18 years.

Other causes that may present with rather similar features are chronic constrictive pericarditis, retroperitoneal fibrosis and cirrhosis of liver.

Hepatography has been used for the diagnosis of Budd - Chiari syndrome. [2]

ACKNOWLEDGEMENTS

I am grateful to Prof. H.O. Wong for her encouragement and my secretary, Miss Saw, for typing the manuscript.

REFERENCES

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