INTRODUCTION

Budd Chiari Syndrome (BCS) is a rare disorder with prevalence of 2.4 cases per million population according to a Japanese study.1 It is characterized by hepatic venous outflow obstruction anywhere from small hepatic veins to atrio-caval junction regardless of etiology.2 It can be primary, with obstruction originating from within the venous lumen, or secondary, resulting from invasion of extra-luminal lesion like abscess, cyst or tumour. A case of BCS, resulting from renal tumour extension in hepatic vein, is hereby reported.

CASE REPORT

A 52 years old lady was admitted with abdominal pain and distention for 2 weeks and drowsiness for one day. Laboratory investigations revealed anemia, thrombocytopenia, INR 2.26, serum bilirubin 3.3 mg/dl, normal transaminases and serum albumin 2.2 gms/dl. Laboratory workup confirmed the presence of de-compensated liver disease with negative viral serology. Abdominal ultrasound and CT abdomen revealed tumour in right kidney with tumour thrombus extending in inferior vena cava and hepatic vein resulting in Budd Chiari syndrome and ascites. Patient was managed with medications only due to advanced liver disease.

DISCUSSION

The Budd Chiari syndrome that results from obstruction of the suprahepatic venous drainage by the tumour could evolve toward liver fibrosis and death.3 Hepatocellular carcinoma, adrenal tumours and renal cell carcinoma are the major malignant conditions capable of direct IVC invasion and venous obstruction.4 Marshall et al. reported that tumour thrombus, invading inferior vena cava in patient with renal tumour, is seen in 4-10% cases5 but hepatic vein invasion by tumour thrombus in patients of renal cancer is extremely rare.6 Once IVC is obliterated, systemic venous return tries to re-establish its flow to right atrium by developing collaterals. Collaterals drain via vena azygos by means of its extensive communications with the lumbar and renal veins or extensive peri-renal collateral circulation.3 But extension of thrombus in hepatic veins can result in hepatic failure due to inadequate collaterals available for hepatic venous return.

Liver biopsy, showing features of centri-zonal congestion, cell necrosis and hemorrhage is the gold standard for diagnosis of Budd Chiari syndrome,7 but liver biopsy also has its own pitfalls as far as diagnosis is concerned. It has wide range of features and usually findings are heterogeneous and subject to sampling error. Similarly, there is no correlation between...
histological findings and patient’s outcome. Liver biopsy probably does not have a dominant role in decision-making in a patient of BCS as was suggested by Shaked et al. Bolondi et al. concluded that diagnosis of BCS can confidently be made on Doppler study and CT abdomen. Diagnosis in our patient was based on Doppler study and CT abdomen. Biopsy of liver and kidney was not done due to coagulopathy, marked ascites and non-reversible stage of liver disease.

Urgent surgical management is recommended in patients of renal cancer with tumour extension in inferior vena cava. Surgery before developing decompensated liver disease is the only hope for cure. Once patient develops hepatic failure following hepatic vein involvement, outcome is poor. In a case series from Japan, Kume et al. reported 4 cases presenting at their centre over 7 years along with 8 cases reported in international literature. Seven of them had liver failure at the time of diagnosis and surgical treatment was possible in only one patient. A possible option would be liver transplant along with resection of primary tumour and thrombus extraction from IVC as suggested by Ciancio et al.

As features of liver failure like hepatic encephalopathy and ascites were present, surgical resection of kidney tumour without liver transplantation would have been futile. Early diagnosis is mandatory for curative therapy of this fatal condition especially in countries like ours with no facility for liver transplantation.

REFERENCES


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