VOLUME - 10, ISSUE - 11, NOVEMBER - 2021 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Medicine

A CASE OF BUDD CHIARI SYNDROME IN 16 YEARS OLD MALE

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KEYWORDS :

INTRODUCTION

Budd chiari syndrome is an uncommon disorder defined as hepatic vein outflow tract obstruction, which is independent of the level and mechanism of obstruction(except pericardial disease and cardiac cirrhosis and sinusoidal obstruction syndrome)

Primary budd chiari syndrome is defined as obstruction due to predominantly venous process (thrombosis or phlebitis)

Secondary budd chiari syndrome is defined as compression or invasion of hepatic veins and/or inferior vena cava by a lesion that originates outside the vein. (malignancy)

Budd-Chiari syndrome is a congestive hepatopathy caused by blockage of hepatic veins. This syndrome occurs in 1/100 000 in the general population. Hypercoagulable state could be identified in 75% of the patients; more than one etiologic factor may play a role in 25% of the patients.

Two of the hepatic veins must be blocked for clinically evident disease. Liver congestion and hypoxic damage of hepatocytes eventually result in predominantly centrilobular fibrosis. Doppler ultrasonography of the liver should be the initial diagnostic procedure. Hepatic venography is the reference procedure if required. Additionally liver biopsy may be helpful for differential diagnosis. The prognosis of the chronic form is acceptable compared to other chronic liver diseases.

Case presentation

16 years male patient presented with complaint of acute abdominal pain since 1 month , abdominal distension since 1 month, Nausea and vomiting since15 days.He had reported his symptoms had begun 1month ago and gradually worsening, increase abdominal girth with progressive dyspnoea.

Investigation and treatment.

CBC-hb 13.1gm /dl,WBC -8300cells /cumm, platelet -1,13000, MCV -76FL. Platelet are slightly reduced in peripheral smear.

All sera-sugar-64 mg/dl,urea -20mg/dl,s.creat-0.4mg,s.Na^+ -129mEq/L,s.K^+ -4.4 mEq/L,s. Cl^- 99mEq/L, S.billi -1.0 mg/dl, SGPT -37 U/L,Alk phos-192 IU/L.

Pt come with some private report , which are done 10 days ago for jaundice S/0- SGOT-393 IU/L,SGPT-284 IU/L, S. Alk phosphatase-510IU/L, s.billi-1.44mg /dl,s. billi direct -0.7 mg/dl.for this patient taken treatment.

S. protein T-6.0 gm/dl, Alb-2.8, Glb -3.2g/dl
Ultrasonography of abdomen –Liver –enlarged,bright in echotexture,increased pericholecystic and periportal echogenicity.

Urinary bladder-Thickened wall P/O cystitis.

ECG WNL NSR, 2decho –no valvular heart disease, EF -60%
Ascitic Fluid analysis-protien -2.4 gm/dl, albumin -1.33 gm /dl,glucose 163mg/dl, Total cells 310,Lymphocytes 94%, Polymorphs 6% ADA-4.38U/l
SAAG –S.Alb-Ascitic alb=2.8-1.33=1.47gm/L, which is more than 1.1 gm/L and ascetic fluid less than 2.5 gm/L.

Opthalm refer for KF ring – Which is absent in bilateral eye.
S. ceruloplasmin -0.55 gm/L
Urine RM (normal), Urine Culture Sensitivity – No growth

HIV 1 and HIV 2 –Negative, HbsAg-Negative, HCV –Negative, HAV -Negative, HEV -Negative

CECT abdomen findings suggestive of:
Caudate lobe and central areas of liver shows early nodular heterogenous post contrast enhancement as compared to peripheral areas with right hepatic vein thrombosis.

Moderate ascites

P/o Budd Chiari Syndrome appears likely.
Lipid profile
S. cholesterol 140mg/dl
Triglyceride-130 mg /dl
HDL-60mg/dl

Patient was treated with Low molecular weight heparin for 5 days and overlap with warfarrin. Eventually Patient 's abdominal pain decrease , but ascites not relieved.Then gastroenterologist opinion taken and so diagnosis was confirmed for Budd chiari syndrome and Advised for-stenting as well as placement of transjugular intrahepatic shunt in acute form of budd chiari syndrome.So the patient was transferred to the higher centre for stenting and further management.

CONCLUSION

16 years male Patient diagnosed as uncommon disorder – Budd Chiari Syndrome which is idiopathic.

REFERENCE

1. Harrison's principle of Internal medicine, 20th edition