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CASE REPORT

Constrictive Pericarditis Leading to Cardiac Cirrhosis: A Rare Cause of Chronic Liver Disease

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Introduction

The relation between diseased heart and liver may manifests as acute liver injury, chronic congestive hepatopathy, even cardiac cirrhosis. Congestive hepatopathy caused from impaired blood return to the right ventricle with increased filling pressure.[1] Chronic liver disease (CLD) is the most frequent presentation of hepatobiliary disease.[2,3] Neonatal cholestasis and metabolic liver disorders are the leading causes of paediatric CLD, whereas autoimmune hepatitis, chronic hepatitis B, C infections, NAFLD, congenital hepatic fibrosis, BUDD chiary disease is least common.[4] Very rare cause, like long term right heart failure may also be a cause of underlying disease for CLD. Our case will present such a short report or cardio-hepatic relations.

Case report

A 11 years old boy of nonconsanguinous parents immunized as per EPI schedule, admitted at BSMMU at Paediatric Gastroenterology and Nutrition department with the complaints of gradual abdominal distention since 6 years of age, more marked for last 2 weeks and legs swelling, respiratory distress for last 2 months associated with exertional dyspnea, orthopnea and nonproductive cough. He also developed jaundice for last 1 month. Patient gave the history of osteomyelitis at 4 years of his age, operation needed two times four months apart for osteomyelitis followed by pericardiocentasis also needed along with injectable antibiotics for 6 weeks. Then he developed anasarca and was treated with oral diuretic (furosemide) off and on. Developmentally he is age appropriated. He has no family history of liver disease, no previous history of jaundice, fever, bleeding manifestations or contact with tubercular patient.

On general physical examination he was cooperative, dyspnoeic, afebrile, moderately pale, mildly icteric, engorged neck veins present, pulse-116 beats/min, low volume, BP-100/60 mm of Hg, BCG scar present. His right arm was deformed with a scar mark. Stigmata of CLD (thenarar-hypothenar wasting, leukonychia) and pedal oedema present. Bed side urine for albumin was nil. He was severely undernourished, severely stunted, mildly wasted. On

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systemic examination, abdomen was hugely distended, flanks were symmetrically full, seen, nontender hepatomegaly (10 cm from the right costal margin, firm with smooth surface.) No splenomegaly, ascites was present. Respiratory system revealed dyspnea, respiratory rate - 44 breaths /min, Spo_2 was 96% in room air. There was Intercostal and subcostal in drawing. No mediastinal shifting. Vocal fremitus was decreased, percussion note dull, breath sound and vocal resonance diminished at both lowest part of chest. Cardiovascular system revealed, location of apex beat on 5th intercostal space at midclavicular line, no palpable heart sound, S_1 S_2 was audible, muffled, no gallop rhythm, no murmur was detected. Neurological system showed no abnormalities.

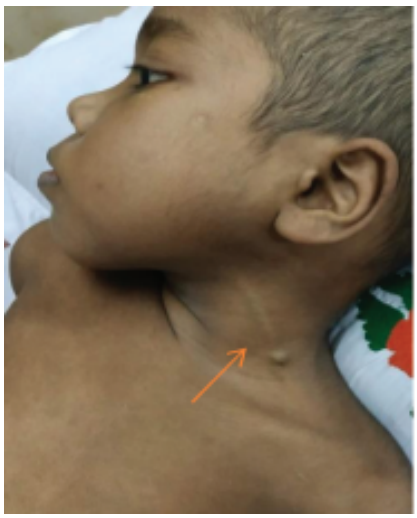

Fig 1 *Engorged neck vein*

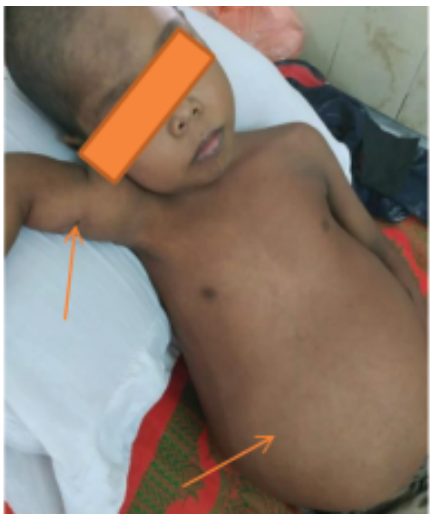

Fig 2 *Deformed right arm with scar mark, huge ascitis*

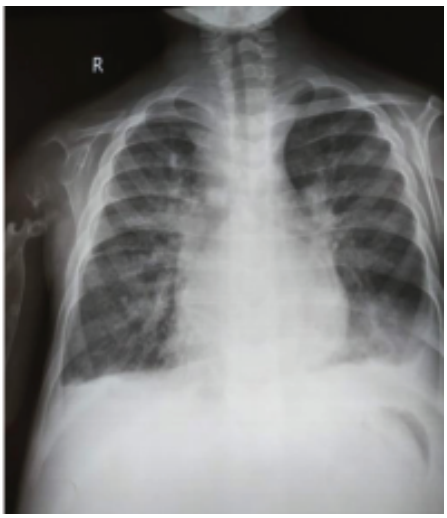

Fig 3 *Bilateral mild pleural effusion, normal heart shape*

Table I
Investigation reports

Investigations	Results	References
Hb% (g/dL)	11	11.5-16
ESR (mm in 1st hour)	15	0- 10
WBC total (/cu mm)	5500	4500-11000
N %	80	40-80
L %	14	20-40
M %	06	2-10
Platelets (/cu mm)	420000	150000-450000
Serum-		
ALT (U/L)	21	35-50
AST (U/L)	76	10-40
Albumin (g/dL)	1.9	3.5-5.0
Alkaline phosphatase (U/L)	150	Upto 300
Ceruloplasmin (mg/dL)	23	20-60
LDH (U/L)	295	208-378
Creatinine (mg/dL)	0.43	0.68
c- GT (U/L)	110	<55
PT (sec)	12	12-16
INR	1	<1.4
Mantoux test (mm)	0	Up to 10

CXR showed bilateral mild pleural effusion with plethoric lung field.

USG of whole abdomen showed moderate hepatomegaly with coarse parenchyma, dilated hepatic vein, huge ascites, no splenomegaly.

Doppler USG of IVC, hepatic vein and portal vein showed features of congestion in IVC and hepatic vein, no evidence of portal hypertension, coarse hepatic parenchyma, huge ascites and bilateral pleural effusion.

Echocardiography showed increased echogenicity and thickness of pericardium dilated right and left atrium, ventricular wall thickness-normal, moderate mitral and mild

tricuspid regurgitations, normal pulmonary atrial pressure, and tissue. Doppler E velocity mildly increased dilated inferior venacava and non-collapsing, diastolic flow reversal during expiration in hepatic vein, suggestive constrictive pericarditis.

Table II
Reports of S. electrolyte, viral markers and ascitic fluid analyusis

Investigations	Results	References
S.Electrolyte		
Na (mmol/L)	127	135-145
K (mmol/L)	4.7	3.5-5.1
CL (mmol/L)	99	95-107
Viral markers		
HBsAg	Negative	
Anti-HCV IgG	Negative	
Ascetic fluid analysis:		
Appearance	Clear	Clear
Biochemical		
protein (gm/dl)	0.7	0.3-4
glucose(mmol/L)	8.2	7-10
Cytological		
WBC (/cumm)	20	<500
N (%)	10	<250
L (%)	90	
malignant cell	00	
Serum ascetic fluid albumin gradient (SAAG) (gm/dl)	1.2	
ADA (U/L)	12.1	1-28

Fibroscan of liver; Median stiffness was 37.4 Kpa, IQR/MED-10%, which correlate with stage-4 fibrosis, that is cirrhosis. Our final diagnosis was cardiac cirrhosis.

Child was kept in bed rest with propped up position and saturation was maintained by oxygen inhalation, salt was restricted in diet with adequate protein supplementation.

Nebulization was given with salbutamol solution and normal saline to relieve bronchoconstriction and breathlessness; 20% Human albumin 1gm/kg along with IV furosemide was given to increase intra vascular compartment volume, to minimize hypoalbuminia and also to reduce the respiratory distress.

According to the cardiac consultation combination of spironolactone and furosemide along with digoxin was given for the treatment of heart failure. Proper cardiac monitoring was done during the hospital stay. During his discharge further follow up plan for both cardiovascular system and also for hepatobiliary system was given. After his discharge from the hospital, he continued to take proton pump inhibitor for the prevention of stress ulcer and lactulose for constipation and also for the prevention of hepatic encephalopathy as per advice.

Discussion

Any type of hepatic fibrosis occurring in cardiac patient is known as cardiac cirrhosis.[5] It is a very uncommon cause of CLD and it's difficult to distinguish from other causes of liver cirrhosis. The most important mechanisms responsible for the development of congestive hepatopathy are hepatic congestion, decreased hepatic blood flow and hypoxemia[6] followed by atrophy, necrosis of hepatocytes, thrombi resulting due to cholestasis.[7]

This case report is one of such scenario of chronic liver injury, leading to fibrosis due to long term congestive heart failure. Causes of cardiac cirrhosis are valvular heart disease, cardiomyopathy, pericardial disease, ischemic heart disease, primary lung disease.[8] Our patient had constrictive pericarditis, bacterial origin, secondary to osteomyelitis. Though in the developing countries tuberculosis is the most common etiology for constrictive pericarditis,[9,10] to support our diagnosis, our patient had negative Monteux test and had no contact history to tubercular patient, Gene Xpart of ascetic fluid was negative. But he had history of osteomyelitis cured by surgery and followed by pericarditis which improved by pricardiocentesis and injectable antibiotics. Subsequently he developed chronic congestive heart failure for 7 years and treated at different hospitals by oral frusemide only Later he developed congestive hepatopathy, menifestrated as jaundice, dyspnea, engorged neck vein, huge hepatomegaly, ascites and pedal oedema, normal S. ALT, alkaline phosphatase levels, raised AST, LDH, bilirubin and low albumin. In congestive hepatopathy, liver function tests do not show the specific pattern as in patient with hypoxic hepatopathy.[11] Cholestatic enzymes together with low albumin and high bilirubin are the strongest risk factor for poor outcome, in case of chronic heart failure.[12] Our patient has high bilirubin and low albumin level. Chest X-ray was suggestive of constrictive pericarditis as there is plueral effusion without significant bilateral enlargement of left and right ventricle. Though calcification may be found in 20%-40% cases of constrictive pericarditis but

more common in tubercular pericarditis.[13] He had also high SAAG (1.1 g/dL) that is transudative.[14] He had no contact history of tubercular patient, Mantoux test, X-Ray chest and ascitic fluid analysis all were negative for tuberculosis. Doppler study shows feature of congestion in IVC and hepatic vein and coarse hepatic parenchyma due to fibrotic changes. Fibro scan by transient elastrography is also now widely recognized as a reliable method to asses liver fibrosis.[15] Though, liver stiffness, shown by transient elastography is not a reliable marker for identifying fibrotic stage of congestive hepatopathy, but it may become a useful non-invasive tool for screening cardiac patients and those who are at risk of cardiac cirrhosis, as increased venous pressure is a risk factor for cardiac cirrhosis[16]. As our patient was suffering from chronic congestive heart failure and ascites, transabdominal liver biopsy is at risk and transjaugular liver biopsy is not practiced at our setting for the evaluation of cirrhosis. So, fibro scan was done and result was suggesting liver cirrhosis. A patient with constrictive pericarditis, develops chronic right heart failure due to markedly elevated ventricular filling pressure, causes passive congestion of hepatic vein, leading to relative ischemia, hepatic necrosis and fibrosis.[17]

Conclusion

Thought constrictive pericarditis and cardiac cirrhosis both are very uncommon, our interest was to highlight the cardiac cause should be evaluated in a dysphonic child, where the causes of CLD were not certain.

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