

An Unusual Case of Cirrhosis Cured by Pericardial Stripping

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Abstract

Congestive hepatopathy refers to hepatic derangement due to venous congestion in right side heart failure. It occurs due to transmission of elevated pressure in right atrium via inferior vena cava and hepatic veins. The aetiology involves most of the causes of right heart failure. Symptoms varies from asymptomatic to symptomatic including feature of right heart failure.

We are presenting a case of 11 year old girl presented with features of progressive decompensated chronic liver disease had amazing improvement after Pericardial stripping.

It is diagnosed with laboratory investigations, including liver functions, imaging including ultrasounds and echocardiography which is an important non-invasive test to evaluate underlying cause of congestive hepatopathy. Treatment is directed towards eliminating the cause and modify effects of the heart failure.

Keywords: Cirrhosis; Pericardial Stripping; Congestive Hepatopathy

Introduction

Congestive hepatopathy refers to hepatic derangement due to venous congestion in right side heart failure.

Case Report

11 year old girl presented with progressive generalized oedema for one week duration associated with exertional dyspnoea (NYHA grade 3). She didn't have any urinary symptoms and bleeding manifestations. She was the second child born to non-consanguineous healthy parents with one healthy sibling.

2 years back she had been admitted with difficulty in breathing and progressive body swelling where she was found to have bilateral pleural effusion and large pericardial effusion which needed aspiration. Since then she was apparently well without repeated admissions.

Her development was normal with average school performance.

Her height is 144 cm, weight 53 kg, BMI 25.5 kgm². Examination showed periorbital oedema, bilateral ankle oedema. She was not icteric, conscious and rational. Abdomen grossly distended with gross ascites. Respiratory system examination revealed reduced air entry on right side lower zone. Central nervous system examination unremarkable. The rest of the physical examination was unremarkable.

Her albumin was 24 g/dl and INR was 1.9. She needed albumin transfusion to settle oedema and then commenced on liver failure regimen. Liver biopsy suggestive of chronic liver parenchymal disease.

She was extensively investigated along the line of chronic liver disease in which the Wilson's disease and autoimmune hepatitis were excluded. Liver sonography shows cirrhosis with moderate ascites. The preoperative laboratory values as well as MELD score presented in table 1. Due to presence of end staged liver disease, patient was admitted for evaluation of liver transplant.

Lab value	Count
Serum creatinine	1.6 mg/dl
Bilirubin	1.8 mg/dl
International normalized ratio (INR)	1.8
Gamma GT	455 U/l
Aspartate aminotransferase (AST)	33 U/l
Alanine aminotransferase (ALT)	22 U/l
Serum sodium	135 mmol/l
Model of end stage liver disease (MELD) score	21 points

Table 1

CECT of abdomen and chest revealed chronic pericardial effusion with dilated inferior vena cava. Focal bulging of right ventricle into pericardial sac? Aneurysm (Figure 3). No hepatic and portal vein thrombosis.

The echocardiography shows congested IVC and hepatic vein (Figure 2) with moderate left and right atrial dilatation. Except for mild tricuspid and mitral regurgitation significant valvular diseases excluded. Bi ventricular systolic functions preserved. There was an abnormal septal movement “septal bouncing” (Figure 1), which is characteristic of constrictive pericarditis.

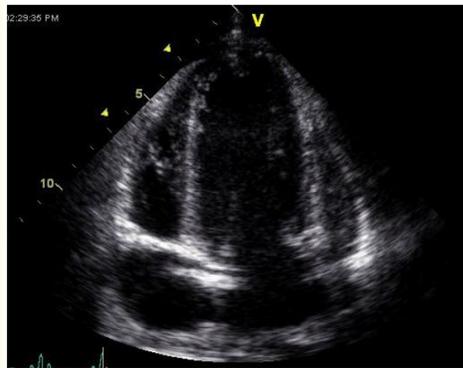


Figure 1



Figure 2



Figure 3

Pericardial stripping with constrictive pericarditis was performed and histology revealed fibrous tissue with no inflammation. There is an area of loose connective tissue with congested blood vessels. Tuberculosis which is the commonest cause for constrictive pericarditis was excluded.

The post-operative period was without major complications. Later the patient fully recovered to NYHA 1 and with a normal liver functions. The laboratory findings at follow up is presented in table 2.

Lab value	Count
Serum creatinine	1.04 mg/dl
Bilirubin	0.8 mg/dl
International normalized ratio (INR)	1.2
Gamma GT	70 U/l
Aspartate aminotransferase (AST)	71 U/l
Alanine aminotransferase (ALT)	30 U/l
Serum sodium	138 mmol/l
Model of end stage liver disease (MELD) score	9 points

Table 2

Abdominal ultrasound scan shows no evidence of cirrhosis and ascites as well. 2D echo shows grade 1 TR and normal mitral valve. Normal LV and RV with preserved systolic as well as diastolic functions were noted.

Child was initially commenced on liver failure regimen but later all the drugs were discontinued.

Discussion

Cardiac cirrhosis also known as congestive hepatopathy even though a well-known disease entity most commonly misdiagnosed. In our case patient’s liver regenerated completely after cardiothoracic intervention. Early postoperative period in patients after, surgical pericardiectomy are clinically challenging, requiring intensive care, invasive hemodynamic monitoring, vasopressor support and diuretics. However, surgical pericardiectomy can be accomplished with excellent midterm outcome [2]. Lin., *et al.* reported 93.7% of patients alive at one year follow up: increased erythrocyte sedimentation rate, impaired renal functions, postoperative low cardiac output and pleural effusion were associated with increased mortality in their sample [1]. But fortunately above complications were not occurred in our patient.

Our case is a good example which clearly shows that constrictive pericarditis should be considered in all the patients with cirrhosis where no cause being found as aetiology. In cryptogenic cirrhosis a cardiac cause should be considered and patients should be evaluated with non-invasive imaging (2D echo, MRI, CT) as well as invasive cardiac catheterization of the hepatic veins with central venous pressure and Trans hepatic pressure measurement in case of failing routine non-invasive cardiological evaluation [3]. So, in this case we were clearly able to avoid liver transplantation.

The cause of liver injury to be evaluated at the time of presentation. Careful history and examination should be done to evaluate possible cause of liver injury like hepatitis, metabolic disorder, medications, gallbladder diseases etc.

Alteration of synthetic functions of the liver presents with prolonged coagulation time. Hypoalbuminemia is rarely less than 2.5 g/dl. The degree of reduction of albumin doesn't correlate with degree of liver damage as this is secondary to bowel oedema from Heart failure leading to protein losing enteropathy and malabsorption.

An echocardiography is an important non-invasive test to evaluate underlying cause of congestive hepatopathy. The echo evaluation needed for valvular heart disease, wall motion anomalies, pulmonary artery pressure or constrictive pericarditis. Inferior vena cava with respiration variation (normally equal or more than 50% of narrowing during inspiration) or IVC diameter above or equal to 2 - 3 cm suggest right heart disease with increase right atrial filling pressure.

The management of cardiac cirrhosis depends on the underlying cardiac condition which causes the hemodynamic imbalance. Adequate fluid management for these patients after assessing the volume status is important.

Pericardial stripping also known as pericardiectomy is the surgical removal of pericardium. Stripping of as much as possible of pericardium is advisable to have the favourable effect of surgery. In most of the patients ascites, peripheral oedema, and congestive hepatopathy would be normalized more quickly.

The take home message at the end of our case presentation is never abandoned good clinical history, physical examination for signs of constrictive pericarditis in a patient with cryptogenic cases of cirrhosis [5]. The history taking together with physical examination and analytical interpretation would lead you to correct diagnosis.

Conclusion

Congestive hepatopathy refers to hepatic derangement due to venous congestion in right side heart failure. It is very important to get a good clinical history, physical examination for signs of constrictive pericarditis in a patient with cryptogenic cases of cirrhosis. The history taking together with physical examination and analytical interpretation would lead you to correct diagnosis.

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