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Research Paper

Cardiac Cirrhosis-Case Report of a Rare Manifestation of a Rare Congenital Heart Disease

Medical Science

KEYWORDS : Congenitally corrected transposition of great arteries, Atrial septal defect, Heart failure, Ascites, Cardiac cirrhosis.

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ABSTRACT

Cirrhosis presenting with ascites is a common condition we come across. Here, we present a case report of 52 year old lady with recurrent episodes of refractory ascites. She was subsequently diagnosed to have cirrhosis of liver & a complete work-up of hepatic causes for cirrhosis was negative. Later we found the signs of heart failure and an underlying congenital heart disease(CHD) i.e, Congenitally Corrected Transposition of Great Arteries(CCTGA) with Atrial Septal Defect(ASD). CCTGA is a rare disease accounting for 0.5-1% of all the congenital heart diseases1, when undiagnosed in childhood, present with symptoms of Right heart failure in their adult life.2 So we report a case of Cardiac cirrhosis secondary to the chronic congestion from right heart failure3 due to an untreated CCTGA with ASD, to highlight this manifestation of congenital heart disease & also to reiterate the cardiac cause as a differential diagnosis when there is Liver Cirrhosis.

INTRODUCTION :

CCTGA means both atrio-ventricular and ventriculo-arterial discordance called double discordance. It is a rare complex congenital heart disease often associated with Ventricular Septal Defect(VSD)-75%, Pulmonic or sub-pulmonic stenosis-75% or Tricuspid valve abnormalities/Ebstein like anomalies in >75% [4], but its association with ASD is not well described in the literature. These conditions cause future complications if left untreated. So early detection & surgical repair should be considered before complications develop.

Cirrhosis of liver, a commonly encountered problem in India, caused by various etio-pathologies. Most common being Alcohol followed by infective & other causes which lead to irreversible damage of the liver parenchyma & land up with its complications if failed to diagnose & manage early. Finally, they will be left with no other option than to join a long waiting list of hepatic transplantations. In the past, it was thought that every cirrhosis was irreversible. But now, it has become apparent that when the underlying insult has been removed, there can be reversal of fibrosis.[5] In such a scenario, it becomes a bigger challenge to identify such a rare cause of liver cirrhosis being a congenital heart disease, which may need an early surgical repair or a Combined liver heart transplantation. As a result of successful reparative surgery for complex congenital heart disease,approximately 85% of patients with CHD now survive into adulthood.[3,6]Hence we present this case of CCTGA with ASD causing Cardiac Cirrhosis as there are a very few such cases reported.

CASE REPORT : A 52 year old female presented with recurrent episodes of abdominal distension, dyspnoea with orthopnea , bilateral ankle oedema from 5 years. No history of abdominal pain/lump/ vomiting/diarrhea. She had no history of decreased urine output/facial puffiness/haematuria/fever. She gave a history of congenital heart disease diagnosed 18 years ago for which she has not undergone any treatment. She denied any history of cyanotic spells / syncopal attacks / Shortness of breath/ palpitations / chest pain/ pedal edema during childhood. she is not a known hypertensive or diabetic. No past history of Tuberculosis/Asthma/Hypothyroidism. No History of Blood transfusions. She was neither a smoker nor an alcoholic. Her Sleep & appetite decreased. Bladder & bowel habits were normal. Without any complications she could complete her family with three normal vaginal deliveries & attained menopause 10 years ago. No History of post menopausal bleeding.

On physical examination, she was Thin built and ill nourished with pallor, bilateral pitting pedal oedema, engorged neck veins and abdominal distension. No other signs of liver cell failure/ clubbing/ cyanosis/ lymphadenopathy. No Kayser-Fleischer ring on slit lamp examination. Her vitals were stable. On examination of the abdomen she had a tense ascites(abdominal girth-156cms) and prominent veins with no obvious organomegaly.

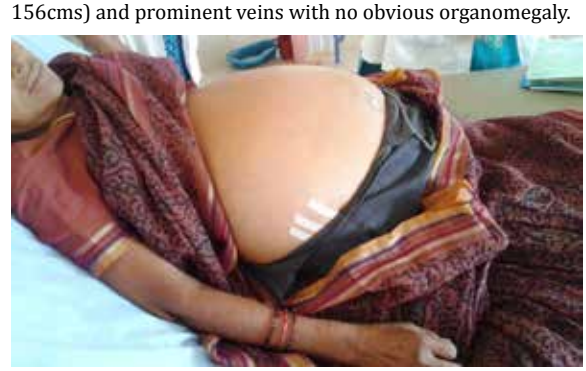

Figure-1: Picture showing the patient with tense ascites.

On cardiovascular examination she had a Raised JVP , Apical impulse displaced 1 cm laterally, Loud 2nd heart sound and Ejection systolic murmur in the left parasternal region in the 2nd and 3rd Intercostal space. On Respiratory system examination , there were diminished breath sounds in bilateral lower zones.CNS examination – No focal neurologic deficits/involuntary movements.

Her Hemogram revealed a mild normocytic normochromic anemia with Hb – 10 g%. Liver function tests showed a normal serum bilirubin and liver enzymes. Serum Total protein – 5.1 g/dl

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, Albumin – 2.2 g/dl. Prothrombin time elevated – INR 2.1. HIV , HBs Ag , HCV – Non reactive. Her Blood sugars, renal parameters, Thyroid profile were within normal limits. Autoimmune markers for liver disease were negative.

Ultrasound Abdomen revealed Mild Hepatomegaly with coarse echotexture and nodular surface with collaterals in falciform ligament, Portal vein normal in dimensions with normal portal vein Doppler study and prominent Inferior venacava , massitive ascites. Biliary tract , Spleen and gall bladder were normal. Ascitic fluid examination revealed Serum ascites albumin gradient(SAAG) - 1.6 g/dl, Suggestive of transudate fluid, with no pus cells/ malignant cells/bacteria, culture was sterile, Adenosine deaminase (ADA) was normal.

Postero-Anterior Chest Radiograph showed an enlarged cardiac silhouette with Right Pulmonary hilum elevated than the left.

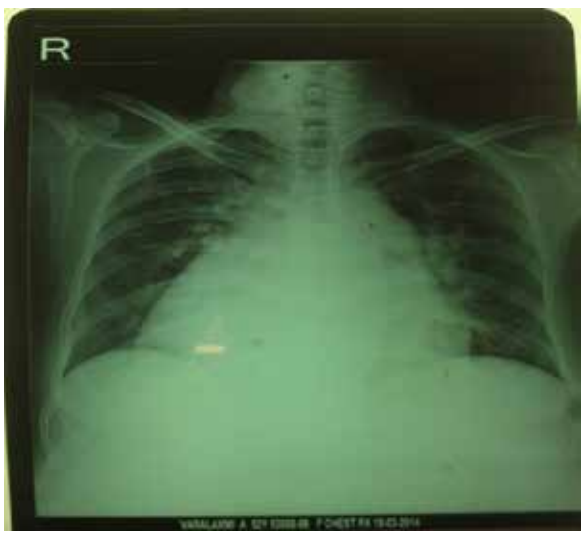

Figure-2: Chest Radiograph showing enlarged cardiac silhouette.

Electrocardigram showed : Normal sinus rhythm with absence of 'Q' waves in left sided chest leads due to abnormal direction of septal depolarization from Right to Left.

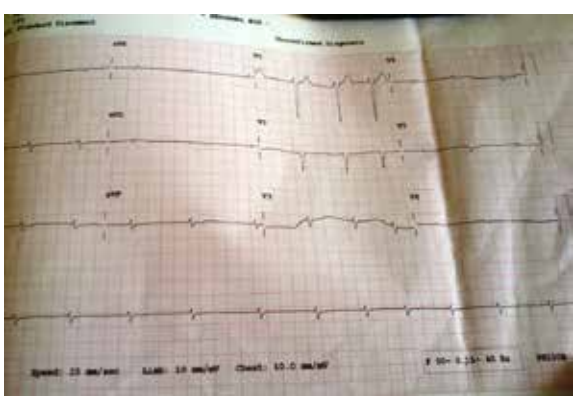

Figure-3: Electrocardiogram showing absent Q waves In left sided chest leads.

Echocardiography revealed: Mitral valve & Left ventricle morphologically on right side & Tricuspid valve&Right ventricle were morphologically on left side. Aorta was Left & Anterior, arising from morphological Right ventricle. Pulmonary artery was right & posterior, arising from morphological left ventricle. A small defect with blood shunting between both the atria was seen. The whole picture, suggesting of Congenitally Corrected Transposition of Great Arteries with Atrial Septal Defect(1cm). There was Severe Systolic dysfunction of ventricles(Right> Left),Dilated chambers,Moderate atrioventricular valve regurgitation on both sides ,Moderate Pulmonary arterial hypertension with Ejection fraction-35%,No pericardial effusion or clots

Figure-4: Echocardiogram picture showing right ventricle opening into aorta & left ventricle into pulmonary artery.

Upper Gastro Intestinal Endoscopy showed No abnormality/ varices.

Liver biopsy, Cardiac magnetic resonance and Cardiac Catheterisation were not done.

Patient was treated with Diuretics(Furosemide 80 mg & Spirinolactone 150mg), Digoxin & Albumin infusion was given. She was advised low-salt, protein-rich diet. Therapeutic paracentesis was done.(fig-5) Inspite of medical therapy & frequent paracentesis, she is only temporarily relieved of her symptoms.

Figure-5: Ascitic fluid

DISCUSSION :

CCTGA is a rare disease accounting for 0.5-1% of all the congenital heart diseases and its association with ASD is very rare[1]. Definition: The term *congenitally corrected transposition of the great arteries* describes hearts in which there are discordant AV connections in combination with discordant ventriculoarterial connections. When there is the usual atrial arrangement, systemic venous blood passes from the right atrium through a mitral valve to a left ventricle and then to the posteriorly located pulmonary artery. Pulmonary venous blood passes from the left atrium through a tricuspid valve to a left-sided right ventricle and then to an anterior, left-sided aorta. The circulation is thus "physiologically" corrected, but the morphologic right ventricle supports the systemic circulation.

CCTGA is often associated with Ventricular Septal Defect(VSD) - 75%, Pulmonic or subpulmonic stenosis-75% or Tricuspid valve abnormalities/Ebstein like anamolies in >75% , but its association with ASD is not well described in the literature.

Patients with no associated defects can be asymptomatic until late adulthood. Progressive systemic (tricuspid) AV valve regur-

gitation and systemic (right) ventricular dysfunction(failure) tend to occur from the fourth decade onward, whereas atrial tachyarrhythmias are more common from the fifth decade onward.[5]

Electrocardiography-Q waves are often present in the right precordial leads and absent in the left. First-degree AV block occurs in about 50%, and complete AV block occurs in up to 25% of patients. Atrial arrhythmias may be seen.

Chest radiography characteristically reveals absence of the normal pulmonary artery segment in favor of a smooth convexity of the left supracardiac border produced by the left-sided ascending aorta. The main pulmonary trunk is medially displaced and absent from the cardiac silhouette; the right pulmonary hilum is often prominent and elevated compared with the left, producing a right-sided "waterfall" appearance.

Echocardiography permits the identification of the basic malformation as well as of any associated anomalies. The major role of Cardiac magnetic resonance in cc-TGA patients is to evaluate the systemic right ventricular volume and ejection fraction, conduit function and AV valve regurgitation. Cardiac catheterization is rarely required, before surgical repair to demonstrate the coronary artery anatomy as well as ventricular end-diastolic and pulmonary artery pressures.

Indications for intervention: If moderate or severe systemic (tricuspid, left) AV valve regurgitation develops, valve replacement should be considered(When EF>45%). When tricuspid regurgitation is associated with poor systemic (right) ventricular function, the double-switch procedure should perhaps be considered, although its role remains controversial.[7] Patients with end-stage symptomatic heart failure should be referred for cardiac transplantation. Pacemaker implantation is usual when complete AV block is present. But Transvenous pacing should be avoided if there are intracardiac shunts because paradoxical emboli may occur.[8]

Therapy:

ACE inhibitor or beta blocker therapy for patients with systemic ventricular dysfunction may be intuitive, but the role of such agents has not yet been demonstrated conclusively.9,10,11. Surgical options being Conduit Replacement or Repair,Tricuspid Valve Replacement,Double-Switch Procedure,Cardiac Transplantation.

Cardiac Cirrhosis : CHD is more common than cardiac disease associated with liver disease. Several CHD defects may lead to either left or right ventricular failure. In these cases, hepatic dysfunction may ensue as a result of the Primary cardiac defect from a combination of passive venous congestion of the liver and hypoxia. Volume overload and low cardiac output may lead to both congestive hepatopathy and hepatic ischemia inturn leading to hepatic fibrosis.12,13.In addition, the presence of cardiac disease and subsequent passive congestion may itself predispose the liver to hepatic injury.14 Over time, cardiac cirrhosis (i.e., central vein to central vein bridging fibrosis and nodule formation) may develop and result in portal hypertension (PH) with ascites and varices.

The most common biochemical abnormalities in passive venous congestion of the liver are elevated indirect bilirubin and prolonged international normalized ratio (INR) with minimal elevations of the aminotransferases. Among patients with chronic liver disease undergoing cardiac surgery , patients with disease of mild severity (Child-Pugh A) did well; high morbidity and mortality were observed in more advanced liver disease.[15]Significant pulmonary hypertension and/or right heart failure may exist in patients with CHD, leading to perioperative hemodynamic instability and thus suboptimal outcomes.[16] Cirrhotic patients have decreased effective circulating arterial volume, which may be further reduced by impaired venous return resulting from tense ascites and diuretic therapy.[17] Postoperatively, low cardiac output may reduce hepatic perfusion, but judicious perioperative support may lead to better outcomes.[16,17.]There are no data to predict outcomes in adult patients with CHD and liver disease undergoing cardiac surgery . The potential for air embolism during the liver transplant procedure(leading to either pulmonary embolism or paradoxical emboli and cerebral infarction) and the risk of infective endocarditis need to be considered.[17,18] In adults, significant cardiopulmonary disease is a relative contraindication to liver transplant and the presence of significant pulmonary hypertension is associated with poor outcomes.[19] There are limited data on combined heart liver transplantation in patients with congenital heart disease.[20,21,22.] The procedure has been performed in selected cases at a handful of centers and may be an option for heart transplant candidates with cirrhosis or for patients with liver failure or HCC secondary to cardiac cirrhosis. Whether earlier cardiac transplantation in patients without cirrhosis can prevent the subsequent development of liver-related complications or impede progression to advanced fibrosis remains unclear.[23]

In case of our patient , as she falls under class –B of Child Pugh's scoring system there is an increased risk of morbidity and mortality to undertake the cardiac surgery. So she is being followed up with medical management and therapeutic paracentesis.

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