CONGENITAL MITRAL STENOSIS - A CASE REPORT

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Abstract:
Isolated congenital mitral stenosis is an extremely rare condition. Congenital mitral stenosis can be due to anyone of the following anatomical variations. These are anamolous mitral arcade, double orifice mitral valve, accessory mitral valve tissue, supravalvular mitral ring, parachute mitral valve and shortened or thickened chordae tendinae. Shone's complex is a condition in which there is multiple obstructive lesions in the left side of the heart and includes Supravalvular mitral ring, Parachute mitral valve, Sub aortic stenosis and Co arctation of aorta. In congenital mitral stenosis most of the patients become symptomatic within two years of age. The patient usually presents with dyspnea, tachypnea, recurrent respiratory tract infections and failure to thrive. Chest X ray will show features of left atrial and right ventricular enlargement. In electrocardiogram evidence of left atrial enlargement (broad P waves in lead II and V1) and right ventricle enlargement (tall R waves in right precordial leads) will be present. Two-dimensional echocardiography with Doppler imaging is the most important diagnostic tool. Surgical correction is the definitive treatment. Here we describe a three years old female child with isolated congenital mitral stenosis. The child was admitted with signs of congestive cardiac failure. On echocardiogram a thin ridge of fibrous tissue attached to mitral valve leaflets was seen and a diagnosis of congenital mitral stenosis due to supravalvular mitral ring was made. The child was treated with antifailure measures and the condition of the child improved with antifailure treatment. The child has been registered in our cardiothoracic department for surgical correction and was awaiting for surgery.

Keyword: supravalvular mitral ring, parachute mitral valve, Shone’s complex

CASE REPORT:
A 3 years old female child was brought to our hospital with breathlessness, cough and cold, fever, pedal edema and abdominal distension for 3 days duration. The breathlessness has been gradually progressing since 1 year of age. The mother also reported that child had recurrent respiratory tract infections and poor...
weight gain since 1 year of age. On physical examination child was found to have pallor, dyspnea and grade 4 malnutrition. She weighed 6 kgs. Her respiratory rate was 52/min. Her heart rate was 142/min and regular. Her pulse was palpable in all the 4 limbs and blood pressure was 85/60 mm Hg. There was no cyanosis or clubbing. Precordial bulge with parasternal heave was present. The first heart sound was loud. P2 was loud and palpable. A diastolic murmur was heard in the mitral area. Bilateral crepitations were heard. Abdomen was distended and liver was palpable 4cm below the right costal margin. A provisional diagnosis of acyanotic congenital heart disease with congestive cardiac failure was made and we proceeded with investigations. The chest X Ray showed cardiomegaly with pulmonary plethora. There was straightening of left border of heart indicating left atrial enlargement and pulmonary artery dilatation as suggested by obliteration of pulmonary bay. Electrocardiogram revealed right axis deviation and broad 'P' waves in lead II indicating left atrial enlargement.

**ECG showing broad 'P' waves in Lead II**

The echocardiogram was diagnostic and showed a thin ridge of fibrous tissue closely adherent to the mitral valve leaflets. Mitral valve area was considerably reduced and measured 1.1cm². Severe pulmonary hypertension was present with a Doppler gradient of 86mmHg. A final diagnosis of isolated congenital mitral stenosis due to supravalvular mitral ring was made.

**Apical 4 chamber view showing enlarged left atrium and supravalvular mitral ring**

The child was treated with oxygen, antibiotics and anti-failure measures. The child’s general condition improved with anti-failure measures and the child has been registered in our cardiothoracic department for resection of supravalvular ring and was awaiting for surgery.
DISCUSSION:
Congenital mitral stenosis is seen in 0.5% of the patients with congenital heart disease \(^1-^3\). Mitral supravalvular ring is a rare congenital heart defect, which was first described by Fisher in 1902 \(^4\). It is characterized by an abnormal ridge of connective tissue on the atrial side of the mitral valve. Often the supravalvular ring may enroach on the orifice of the mitral valve leaflets and restrict their movements. Most patients become symptomatic within 2 years of age. These patients will have dyspnea, tachypnea and nocturnal cough due to pulmonary venous congestion. Frequent respiratory tract infections, poor feeding and failure to thrive are the common symptoms in these patients. Hemoptysis and syncopal attacks can occur. A prominent parasternal heave and loud P2 due to pulmonary hypertension may be present. Unlike acquired mitral stenosis, an opening snap is not common in supravalvular mitral ring. A middiastolic murmur may be audible at the apex, especially in the left lateral decubitus posture and it may exhibit presystolic accentuation. Patients with chronic mitral valve obstruction develop signs of congestive heart failure. Complications of supravalvular mitral ring include pulmonary edema, pulmonary arterial hypertension, atrial arrhythmia, left atrial thrombus, embolic episodes and infective endocarditis. Cerebral venous thrombosis has been described in infants with supravalvular mitral ring. The important differential diagnoses to this condition are cor triatriatum, double orifice mitral valve and acquired mitral stenosis. In cor-triatriatum there is a membrane which divides the left atrium into two chambers.

IMAGING STUDIES: In chest radiograph the most common abnormality is left atrial enlargement and is diagnosed by straightening of the left cardiac border (mitralization), widening of the tracheal carina and elevation of the left bronchus. In severe cases, alveolar edema produces a hazy appearance in the hilar regions of both lung fields. Two-dimensional echocardiography with Doppler imaging is the most important diagnostic tool. It depicts the lesion and helps in quantifying the severity of the obstruction. A thin membrane or a ridge of fibrous tissue closely adherent to the valvular leaflets restricting the movements of mitral valve leaflets may be seen. The present case had similar picture in echocardiography where a ridge of fibrous tissue was seen adherent to the mitral valve leaflets restricting its movement. In children, transesophageal echocardiography is generally not necessary because adequate information can be obtained from transthoracic windows itself. Cardiac catheterization and angiography is not needed if echocardiography provides all the necessary anatomic and hemodynamic data.

MANAGEMENT:
Medical therapy helps to relieve symptoms of congestive heart failure but does not correct the underlying anatomical problem. Surgical resection of the supravalvular ring is the treatment of choice. Any abnormalities in the mitral valve should be simultaneously repaired. If the mitral valve apparatus is grossly abnormal then mitral valve replacement should be done. Percutaneous transcatheter balloon dilation has been done in few cases but the results are less satisfactory.

PROGNOSIS:
Among patients with congenital mitral stenosis, those with supravalvular mitral ring have a relatively good prognosis. The presence of a normal underlying mitral valve and absence of other major cardiac lesions are associated with a better surgical outcome.
Recurrent supravalvular mitral stenosis is a risk in as many as 15% of survivors, probably because of continuous turbulence across the small left ventricular inflow tract

REFERENCES:

