Indian Journal of Basic and Applied Medical Research; December 2014: Vol.-4, Issue- 1, P. 23-27

Case Report:

Shone's complex – a rare case report

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Abstract:

SHONE'S COMPLEX is a rare cardiac anomaly consisting of four obstructive lesions of the left heart: supra mitral membrane, parachute mitral valve, subaortic stenosis, coarctation of aorta. We report a 2 year old female child who was initially diagnosed as having aortic stenosis but continued having breathlessness despite being treated with diuretics and beta blockers. She was brought to us in CCF and we diagnosed her to be a case of coarctation of aorta due to absent lower limb pulsations. 2D Echo further elaborated the condition as being Shone's Complex.

Key words: Shone's complex, supramitral membrane, parachute mitral valve, subaortic stenosis

Case report:

A 2 year old female child was brought to us with complaints of breathlessness and 3-4 episodes of seizures since last few days. The child was a diagnosed case of aortic stenosis with bicuspid aortic valve. She had dyspnea at about 1 year of age when she was diagnosed as case of aortic stenosis with bicuspid aortic valve. Child was put on diuretics and beta blockers at I year of age. The child was clinically stable till about 6 months back when she started having 3-4 episodes of seizures per day intermittently. The seizures were classically generalized tonic clonic with uprolling of eyeballs. The parents brought the child to the neurology clinic in our hospital wherein she was found to have absent lower limb pulsations. The upper limb pulsations were preserved. The child had grade II hypertension in the upper limbs and blood pressure wasn't recordable in the lower limb. Child was dyspneic with basal crepitations and had a markedly palpable liver. On examination, the pulses were absent in both lower limbs while the pulse rate was 124/min in the upper limbs. The BP was 110/70 mm Hg in the right upper limb. There was a heaving apex beat in the 6th intercostal area in the anterior axillary line. The 1st heart sound was loud with a mid-diastolic murmur of grade III/IV at the apex. There was a long ejection systolic murmur (grade III/VI) at the aortic area which was conducted. Echocardiography showed characteristic parachute-like mitral valve and supravalvular mitral ring with a severe mitral stenosis (mitral valve size – 1.1 cm2, PG- 13 mm Hg) causing obstruction to flow. Aortic valve was calcific and bicuspid with a moderate aortic stenosis (PG- 40 mm Hg). There was left ventricular hypertrophy with a normal global systolic function and a normal ejection fraction. Above findings were suggestive of SHONE'S COMPLEX. Parents were counseled

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about the condition. MRI Brain was normal. MRI brain with MR angiography was also done which ruled out any associated aneurysm. Child was stable for 2 days and we planned to refer the child to a higher center for urgent surgical intervention. Unfortunately a day prior to referral, child had sudden cardiac arrest and expired.

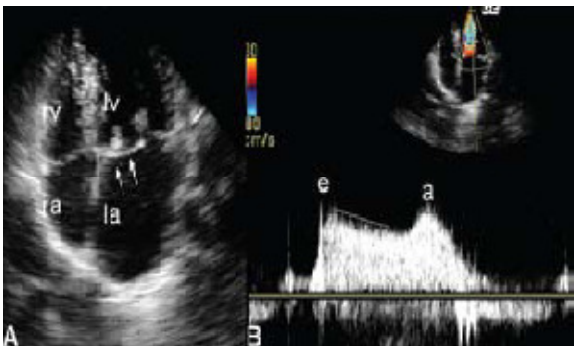


Image 1:

Congenital mitral stenosis Transthoracic echocardiograms.

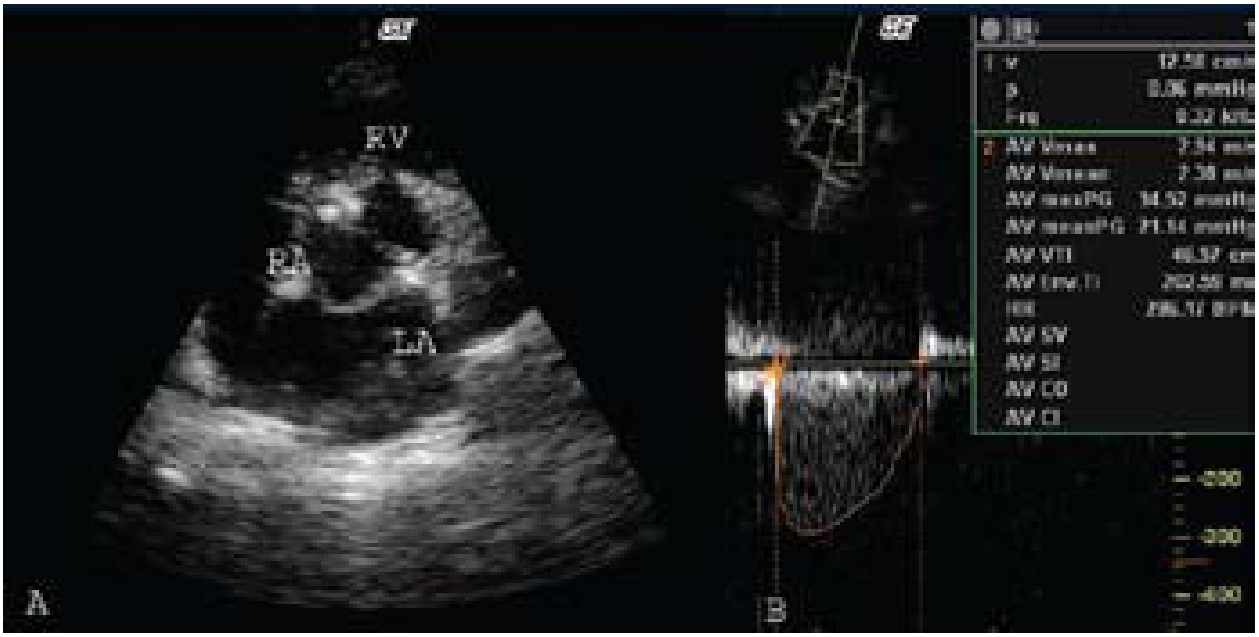
A: Apical four-chamber view. This diastolic frame shows a dilated LA and supravalvar mitral stenosing ring (arrows) that is adherent to the mitral valve.

B: Continuous –wave Doppler studies demonstrated increased peak early (e) and late atrial (a) diastolic flow velocities and decreased diastolic slope (line), peak a-wave velocity is increased, 2 m/s.

Image 2:

Parasternal short-axis view of a bicuspid aortic valve. In diastole the aortic valve is thick. The cusp fusion line and forming the raphe suggests a bicuspid aortic valve.

B: Continuous-wave Doppler signal from the apical four-chamber view shows aortic valve stenosis. The peak velocity is 2.9 m/s. RA-right atrium: LA-left atrium : RV-right ventricle



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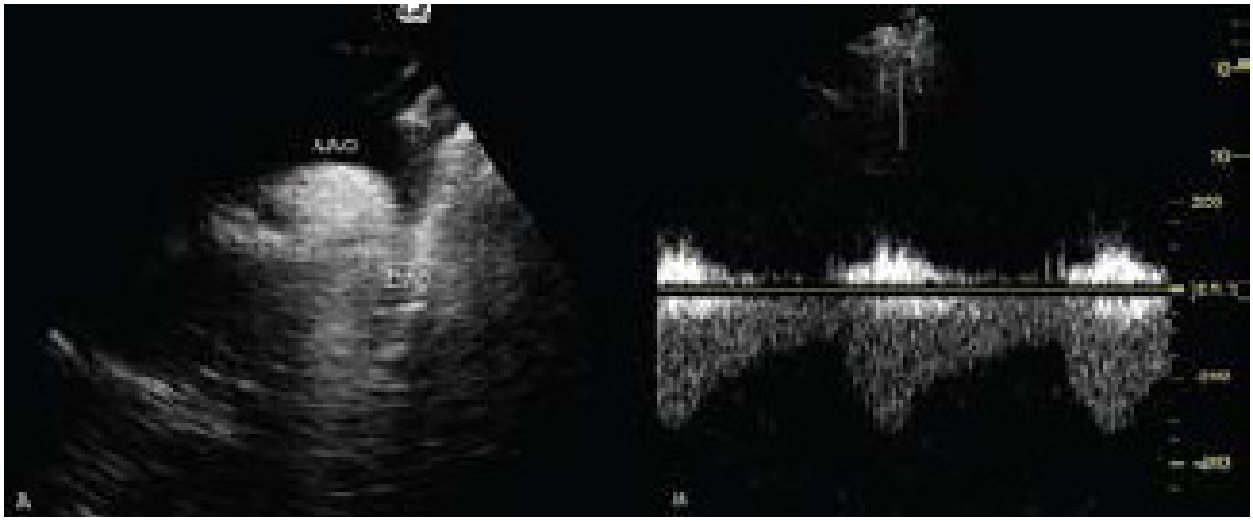

Image 3:

A: Two dimensional echocardiogram obtained in the suprasternal long-axis view shows coarctation of the aorta. B: Typical continuous –wave Doppler display across a severe coarctation. The Peak velocity is 3.2 m/s.

Discussion

Shone's complex is a rare congenital heart disease described by Shone et al initially in 1963. It typically consists of four obstructive lesions of the left side of the heart and circulation namely parachute like mitral valve, supravalvar mitral ring, subaortic stenosis , and coarctation of aorta.1 There is a complete form of Shone's complex wherein all the four lesions are present; however incomplete forms with two or three lesions are also described.1 Other coexisting mitral valve anomalies have been reported such as fused chordae, single papillary muscle and"typical" (Ruckman & Van Praagh) congenital mitral stenosis.2 The LVOT obstruction features may include subaortic stenosis, valvar aortic stenosis, bicuspid aortic valve, and coarctation of aorta.2

Supravalvar mitral ring is a circumferential ridge or membrane, which arises from the left atrial wall overlying the mitral valve and is frequently attached to the mitral valve. The ring may range from a thin membrane to a thick discrete fibrous ridge. It may vary in its extent. Adhesion to the valve may impair opening of the leaflets causing mitral-valve inflow obstruction in some patients.3 In other patients, the ring may be large and protrude into the mitral-valve inflow thus causing obstruction. Parachute mitral valve is defined as a unifocal attachment of mitral valve chordae independent of the number of papillary muscles. A true parachute mitral valve (PMV) is characterized by attachment of the chordae to a single or fused papillary muscle; however PMV also includes asymmetrical mitral valves having two papillary muscles, one of which is dominant and elongated,with its tip reaching to the valve leaflets. The unifocal attachment of the chordae results in a restricted valve opening and subvalvar obstruction and, rarely, valvar regurgitation.4 Oosthoek et al4 suggested that these morphological features distinguish a parachute-like mitral valve from a true PMV. Shone's complex is a rare congenital anomaly. Fewer than 100 patients have been reported in the literature.3 It is mostly detected in childhood as the patient becomes symptomatic by the age of 2 years.3 The usual symptoms are dyspnea, nocturnal cough, tachypnea, poor feeding, failure to thrive, fatigue, and signs and symptoms of heart failure and reduced cardiac output. The child usually has recurrent episodes of wheezing and respiratory tract infections due to pulmonary congestion and exudation of fluid into the lungs.3 The patient may occasionally present with acute pulmonary edema. It is extremely unusual for a patient to remain largely asymptomatic throughout childhood and get incidentally detected during adulthood while evaluating for some unrelated illness. The present case was misdiagnosed as only aortic stenosis with bicuspid aortic valve during 2d echo done at a private setup a year back. Child had absent lower limb pulsations which revealed the presence of a coarctation of aorta.The patient presented to us with congestive cardiac failure and absent lower limb pulsations. This prompted us to investigate the patient in detail. The echocardiographic findings revealed the features of complete form of Shone's complex. A literature search revealed a few articles mostly case reports. Goswami et al5 reported Shone's anomaly in a young gravid female mimicking preeclampsia at 25 weeks gestation. Most of the other reports are in children. Most of these reports are from foreign literature. To the best of our knowledge the present case report is a rare case report of Shone's anomaly from India.A good outcome is possible in patients with Shone's complex,provided the surgical intervention is undertaken early before the onset of pulmonary hypertension.6 Mitral valve repair along with resection of supramitral ring is preferable over valve replacement. Other surgical procedures depend upon existence of associated cardiac anomalies, which ultimately define late surgical outcome. The above patient presented late to us and could have been saved with early diagnosis and early surgical intervention. Unfortunately our patient couldn't be saved but we were able to diagnose this condition in our rural setup was really a feather in our cap.

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