**Shone’s Complex**

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**Abstract**

Shone's complex is a rare congenital heart disease consisting of multiple levels of left sided obstructive lesions including supravalvar mitral ring, parachute mitral valve, subaortic stenosis, and coarctation of aorta. The present case report describes a case of complete form of Shone's complex that was incidentally detected in an adult.

**Introduction**

Shone’s complex is a congenital heart disease, consisting of multiple levels of left sided obstructive lesions including supravalvar mitral ring, parachute mitral valve, subaortic stenosis, and coarctation of aorta.1 This is a very rare malformation and a very few cases have been reported in literature.

**Case Report**

A 30 year old man was admitted to our institution, a tertiary care institution in our state, as a case of pyogenic meningitis and was being treated for the same. On examination, the pulses were absent in both lower limbs while the pulse rate was 64/min in the upper limbs. The BP was 160/110 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 to the carotids.

Based on the above findings, a provisional diagnosis of coarctation of aorta with left sided obstructive valvular lesions in a case of pyogenic meningitis was made.

Various investigations were performed to confirm the diagnosis. ECG showed the left ventricular hypertrophy. On the chest X-ray there was a normal cardiac silhouette and a normal lung parenchyma but there was notching of the lower margins of 6th, 7th, 8th, 9th ribs.

He was subjected to 2-D transthoracic echocardiography (TTE) as well as transesophageal (TEE) echocardiography. TTE showed a severe post-ductal coarctation of aorta (PG-60 mm Hg) with a narrow and tortuous long segment. There was a parachute-like mitral valve and supravalvular mitral ring with a severe mitral stenosis (mitral valve size – 1.1 cm², PG-25 mm Hg) causing obstruction to flow. Aortic valve was calcific and bicuspid with a moderate aortic stenosis (PG-40 mm Hg). There was mild left ventricular hypertrophy with a normal global systolic function and a normal ejection fraction. TEE confirmed the findings of TTE.

The patient was subjected to CT angiography which revealed the location and the size of aortic coarctation. MRI brain with MR angiography was also done which ruled out any associated aneurysm.

The patient was retrospectively probed in detail for the presence of any cardiovascular symptoms in childhood. Though the patient admitted that he used to have dyspnoea of NYHA class I, he was otherwise asymptomatic and had not sought medical consultation earlier.

The patient was treated successfully for pyogenic meningitis. Following this, the patient improved and is now advised...
corrective surgery for which he is referred to a cardiac interventional center (as the facility for cardiac surgery is not available in our institution).

**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. 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. Other coexisting mitral valve anomalies have been reported such as fused chordae, single papillary muscle and “typical” (Ruckman & Van Praagh) congenital mitral stenosis. The LVOT obstruction features may include subaortic stenosis, valvar aortic stenosis, bicuspid aortic valve, and coarctation of aorta.

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 of the valve may impair opening of the leaflets causing mitral-valve inflow obstruction in some patients. 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. Oosthoek et al 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. It is mostly detected in childhood as the patient becomes symptomatic by the age of 2 years. 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. 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 therefore assumes significance. This patient had been largely asymptomatic (except for NYHA class I dyspnea which he had ignored) in childhood. He had presented to us for meningitis during which his clinical examination revealed evidence of mitral stenosis along with left ventricular outflow tract obstruction and aortic coarctation. 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 al 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 the first 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. 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.

**References**


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