Commentary: Shone complex.

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Accepted on August 20, 2018

Commentary

In a recent publication [1], we have reported on a case of Shone complex in an adult lady. Due to extreme rarity of the condition particularly in adult population; we would like to write this commentary. Congenital heart diseases (CHD) are diagnosed in about 1% of all newborn babies. There is an amazing improvement in outcomes for children born with CHD making this one of the success stories of modern medicine [2]. While some of these malformations such as atrial and ventricular septal defects are very common, others like the one described herein are extremely rare. Dr. John D. Shone and his colleagues in 1963 described for the first time a very rare CHD which was later on named after him [3]. Four congenital obstructive or potentially-obstructive left-sided lesions compose the complex; namely aortic coarctation, parachute mitral valve (PMV), supravalvular mitral ring and subaortic stenosis [4]. Worthy to note that Tetralogy of Fallot (TOF) the commonest cyanotic CHD also consists of 4 lesions but exists on the right side of the heart [2].

The malformation of PMV refers to a bi-leaflet mitral valve (MV) with 2 commissures in which the chordae join together to insert into one chief papillary muscle instead of two [4]. The supravalvular ring of the left atrium (LA) is a circumferential ridge of connective tissue that arises at the base of the atrial surfaces of the mitral leaflets and protrudes into the MV inlet. Two types of subaortic stenosis were originally described (muscular and membranous) [4,5].

Our patient was 33-year old lady from Sulaimaniyah, Region of Kurdistan, Iraq admitted to Faruk Medical City (FMC) in July 2014 because of worsening effort dyspnea while she was pregnant. Physical examination was unremarkable apart from cardiac murmur. She had had left thoracotomy for repair of aortic coarctation at the age of 5. Echocardiography confirmed the presence of PMV and aortic stenosis. However, neither supravalvular mitral ring nor residual coarctation of aorta was detected. Open heart surgery was then performed in which the MV was replaced by a prosthetic valve (St Jude 27 mm), subaortic membrane was resected and left ventricular myectomy was done. The outcome was excellent [1].

The presented case is considered incomplete due to the absence of supra-valvular mitral ring. St. Louis et al had only 2 cases of complete Shone complex out of 28 cases (7.1%) [6]. In contrast, Bolling et al. [7] described a complete form of the syndrome in 19/28 (68%) of their patients.

The diagnosis of Shone complex is basically made on clinical grounds and must be confirmed by echocardiography and/or magnetic resonance imaging (MRI) [4]. Although most cases are detected at childhood [5], late diagnosis at adulthood has been infrequently reported [3,4].

The choice of a suitable operative therapy for Shone complex remains difficult. This is due to different presentation and severity of individual lesions [6]. Most reported patients required multiple operations [7]. In one study, 17/27 patients (63%) underwent 2 operations [6].

The prognosis of the treated patients depends on 2 factors; the severity of MV obstruction [7] and the timing of surgery. A good outcome in anticipated in those operated upon prior to the onset of pulmonary arterial hypertension [5].

To sum up, this is a very rare case of incomplete Shone complex in an adult. It is extremely unusual for a patient to remain largely asymptomatic throughout childhood and get incidentally detected during adulthood. Although the definite diagnosis of this case wasn't made at childhood, early surgical therapy of aortic coarctation could have contributed to her negligible symptoms. The patient was successfully managed by conventional valve surgery. Surgery of Shone complex is expected to produce good outcome if performed before the beginning of pulmonary hypertension.

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