Importance of Early Detection of Subaortic Obstruction

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CASE I

12 years old child presented with history of fever and general bodyache and was diagnosed as a case of acute rheumatic fever with carditis, in a medical college, as the child had a systolic murmur and tachycardia. The child was put on steroids and as the condition deteriorated the child was shifted to NIMHANS for history of convulsions and was diagnosed to have tubercular meningitis and treated appropriately. The child was referred to us for cardiac evaluation. 2D echo showed subaortic membrane (Fig. 1) but with no evidence of rheumatic heart disease.

The murmur caused by the subaortic membrane was mistaken for Carey Coomb's murmur of rheumatic carditis! The clinical assessment is important; the murmur caused by subaortic membrane does not fit into ejection systolic murmur of aortic stenosis (AS) or pansystolic murmur of mitral regurgitation (MR) or long systolic murmur of VSD. Hence, echo is the best diagnostic tool which can delineate the type of subaortic obstruction and give information about gradient across left ventricular outflow tract (LVOT), LV function, LV hypertrophy (LVH), aortic regurgitation and associated anomalies. Echo not only helps in accurate diagnosis but also assists in management strategy. Hence, it is very important for every echocardiographer to know about this condition to prevent mistakes like in the above case.

CONGENITAL SUBAORTIC OBSTRUCTION

It is a challenge for the clinician. Echocardiography can help in the precise diagnosis so that appropriate treatment modality can be decided. Subaortic stenosis is divided into three types: 1) Discrete membranous "diaphragmatic", 2) Fibromuscular ridge and 3) Tunnel like obstruction.

Discrete Subaortic Membrane

It is usually associated with other congenital anomalies in 60-70% of cases e.g. VSD, bicuspid AV, coarctation of aorta (COA), atrioventricular septal defects (AV-Canal). The subaortic obstruction is almost always progressive and is not static. Subaortic obstruction may have a "diaphragm" beneath the aortic valve, which may be crescentic or form a complete circle. Often it may be attached to anterior mitral leaflet and this crucial information should be reported so that the surgeon can take precaution during excision of the membrane and avoid damage to anterior mitral leaflet (AML). Because of the likelihood of progressive damage to the aortic valve leading to thickening, regurgitation and sometimes severe LV dysfunction, there is some consensus that the membrane should be removed early to prevent progressive aortic valve damage. There is some debate about whether this lesion is indeed congenital, since it is rarely observed in newborns. It is therefore often regarded as an acquired lesion. It has been proposed that a left ventricular outflow malformation characterized by a wider mitral-aortic separation, an exaggerated aortic override and a steeper aortoseptal angle is present in children with VSD or COA who subsequently develop subaortic stenosis. Clinically it is difficult to diagnose subaortic membrane. Echocardiographically it may be missed because of its close proximity to the aortic valve. One has to look carefully for the fibro-elastic membrane just below the aortic valve. Otherwise one ends up reporting wrongly as just LVH. Though on echo it looks like small ridges, on necropsy they are much more extensive. Therefore careful parasternal long axis imaging as well as apical four-chamber and five-chamber imaging may delineate the defect or sometimes transoesophageal echocardiography may be necessary. Early systolic closure of the aortic valve in M-mode gives a clue to subaortic membrane. As treatment consists of complete resection of the membrane along with a limited myomectomy, it is important for an echocardiographer to evaluate the extent of the diaphragm.

Fibromuscular Subaortic Ridge

It is associated with fibrous ring associated with muscular hypertrophy and located approximately 1 cm below the aortic valve and extending downwards 1-2 cms causing more diffuse area of left ventricular outflow obstruction (Fig. 2 a and b) but also frequently encroaches on the AML. Apical five-chamber view may be useful adjunct, because it places the membrane or ridge perpendicular to the path of the scan plane, thereby enhancing the visualization. Doppler study to obtain the peak velocity is very important and must be distinguished from the signal of VSD and mitral regurgitation (MR).

Subvalvular Fibromuscular Collar or Tunnel

It **p**roduces a more extensive area of obstruction which is characterized by an inward bowing of the echoes from the anterior and posterior margins of the outflow tract immediately beneath the aortic valve. There is more diffuse obstruction extending well into the ventricle associated with left ventricular hypertrophy. In

Fig. 1: Subaortic membrane in parasternal long axis view

Fig. 2b: Subaortic fibromuscular ridge in apical five-chamber view.

Fig. 2a: Subaortic fibromuscular ridge in parasternal long axis view

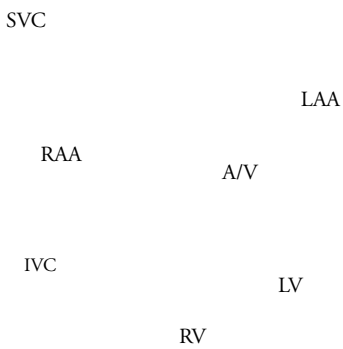

Fig. 3: Diagrammatic picture of cor-triatriatum sinistra

this type there is a dense fibroelastic endocardial tissue involving the entire LVOT, annular hypoplasia and, fibrous cusps and is a major surgical challenge. Therefore, careful imaging in the parasternal long axis and short axis is important to delineate the true dimensions of the LVOT.

Message

The hypothesis that discrete subaortic stenosis is a progressive disorder and may develop into tunnel-like subaortic stenosis

Fig. 4: Echo picture of cor-triatriatum sinistra

as documented by serial hemodynamic and angiographic investigations. One should not allow it to progress to type II or III, as extensive myomectomy could lead to complete heart block in Type II or extensive resection with aortic valve and /or root replacement is needed in type III. Early detection is very important. Hence the onus lies on echocardiographer to detect the subaortic membrane in time when it can be treated with simple excision.

Importance of Segmental Approach to Diagnosis of Rare Combination of Complex Congenital Anomalies

CASE II

18 months male child presented with history of repeated respiratory infections, breath-holding spells, excessive sweating and feeding difficulties. O/E grade III malnourishment, weight 6.5 kgs (Expected wt. - 12 kgs), mild cyanosis and BP - 96/30 mm of Hg. First admission was at the age of 3 months. Detailed echo and doppler evaluation by segmental approach revealed congenital cyanotic heart disease, situs solitus, ASD, cor-triatriatum, TAPVC and PDA with PH. For the understanding of congenital cardiac lesions the 'segmental approach' is recommended. This means that one identifies echocardiographically each cardiac segment independently i.e. atria, ventricles, great arteries and veins and then define their connections and relations methodically. The morphological right atrium is identified by the entry of inferior and superior caval veins and the characteristic appearance of the

appendage (broad and blunt). The left atrium is identified by the narrow and curved appendage. Entry of pulmonary veins to the left atrium is taken as criteria for identification, but is not constant as in case of TAPVC as in this case, they were draining into superior vena cava. One should not be content with identifying just ASD, but look for other associated lesions. Otherwise rare anomalies like cor-triatriatum will be missed.

COR-TRIATRIATUM

Cor-triatriatum sinistra

It is a rare cardiac malformation characterized by the presence of a fibromuscular membrane that divides the left atrium into a distal chamber (DC), which is the true left atrium and is related to the left atrial appendage (LAA) and the mitral valve and the proximal chamber (PC) or accessory left atrium which is related to the pulmonary veins. (Fig. 3) When this membrane is obstructive type the patient presents clinically like mitral stenosis or Leutembacher's syndrome. In its classic form, the accessory chamber (PC) that receives blood from pulmonary veins and communication with left atrium is accomplished by way of one or more fenestrations in the membrane. The malformation is usually isolated but in about one in four patients is associated with other congenital defects of a complex nature. The transthoracic echo (TTE) with Doppler is an excellent modality to diagnose this condition (Fig. 4). Rarely transoesophageal echo (TEE) can give better yield on the number and location of openings in the membrane. This condition should be differentiated from supravalvar mitral ring, which is characterized by the presence of a membrane in left atrium immediately above the mitral valve and does not divide the left atrium into two chambers. The mitral annulus usually appears hypoplastic and the membrane often appears to be incorporated into the mitral leaflets and hence often overlooked. Echo with Doppler is considered as an excellent means of describing all aspects that point to diagnosis, pathophysiology and corrective surgery.

Cor-Triatriatum Dexter

This is an extremely rare anomaly characterized by a membrane that divides the right atrium into two chambers. A distal chamber is related to the right atrial appendage (RAA) and tricuspid valve and the proximal chamber is related to superior and inferior vena cavae and coronary sinus. Rarely membrane could be stenotic. To differentiate it from eustachian valve, the coronary sinus is medial to membrane in cor-triatriatum dexter.

Message

The cross-sectional echocardiography with segmental approach makes the understanding and diagnosis of complex cardiac lesions easy and accurate. The eyes will not see what the mind does not know; hence knowledge about rare anomalies is essential. As the Echo report is like a road map for the management, it is important that the lesions are identified properly, in this direction the segmental approach is very important.