Case Report
Pak Armed Forces Med J 2016; 66(Suppl):S230-32

A CASE OF HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY MISDIAGNOSED AS AORTIC STENOSIS

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Keywords: Aortic stenosis, Calcification hypertrophic Obstructive Cardiomyopathy.

INTRODUCTION

Aortic stenosis results from thickening, calcification and/ or fusion of aortic valve leaflets thus producing an obstruction to LV outflow. The severity of AS can readily and accurately be diagnosed by either echocardiography or cardiac catheterization. Transthoracic echo is most commonly employed for this purpose, being non invasive and reasonably accurate. It not only allows one to gauge the thickness of and calcification over the aortic valve, but helps to measure the severity of AS by such means as planimetry and measuring pressure gradient across the valve. However, gradient can also arise as a result of narrowing of LVOT or systolic anterior motion (SAM) of mitral valve as happens in HOCM. We present a patient who was planned to undergo aortic valve replacement surgery for severe AS, but was discovered to be suffering from HOCM producing AS like symptoms and a structurally normal aortic valve during perioperative transesophageal echo (TOE).

CASE REPORT

A 57 year old male presented with 2 years history of generalized body weakness, weight loss and breathlessness NYHA III. His symptoms were progressive in nature and aggravated particularly over the last 1 month. There was a single episode of blackout as well from which the patient recovered spontaneously. On examination there was a typical crescendo decrescendo murmur best heard along the right sterna edge and radiating to the carotids[2]. D echo showed good overall LV function with normal dimensions and an EF of 60%. There was severe aortic stenosis with PPG of 85mmHg and MPG of 47mmHg. Angiography was unremarkable. One of the reports also showed suspicion of MR II but next one negated the same. Based on clinical findings and investigations, patient was diagnosed to be suffering from severe AS and aortic valve replacement was offered to which he agreed.

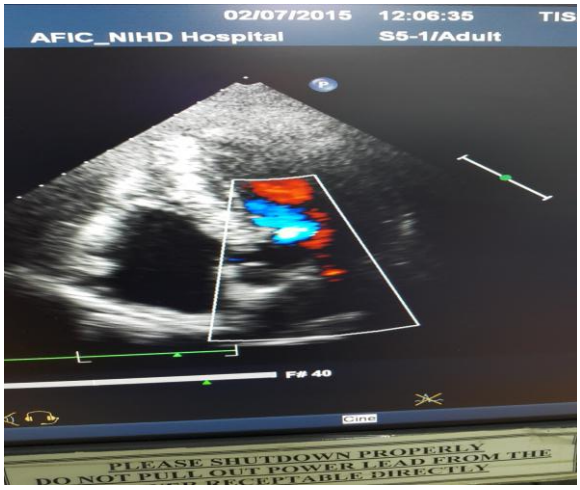

Figure: Two-dimensional echocardiogram in apical four-chamber view.

During the course of surgery however, transesophageal echo showed a little different picture. We noticed turbulent flow in LVOT, where the turbulence appeared to be originating before the aortic valve. Same was confirmed in multiple views. Aortic valve also appeared to be pretty normal with AVA of 2.3cm[2]. There was no calcification of the valve and there was no AR. Due to poor alignment of ultrasonic beam, we could not determine transaortic PG or the gradient across LVOT.

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However, the turbulence was clearly coming before the aortic valve. Septum was very thick and anterior wall of mitral valve seemed to be touching the septum earlier on during systole and leading to narrowing of LVOT, thus giving rise to aortic stenosis type picture. The surgery went on and on opening aorta, our suspicion was confirmed. Aortic valve itself was free of any disease and there was no AR. Interventricular septum was very thick. The surgeon did a wedge resection of the septum and closed aorta. Patient was weaned off CPB smoothly without any inotropic support and was discharged from ITC the very next day. On post operative interview on 5th post op day, the patient felt alright with partial relief of the symptoms.

DISCUSSION

Aortic stenosis results from narrowing of the aortic valve. Underlying pathology may vary from congenitally bicuspid valve, to it becoming calcified secondary to rheumatic heart disease. This result in pressure overload and a symmetric and concentric hypertrophy of the LV. Hypertrophic obstructive cardiomyopathy (HOCM) on the other hand can have genetic basis or can be idiopathic. However, instead of causing symmetric hypertrophy of the LV, it causes asymmetric hypertrophy. In addition, the normal alignment of muscle cells is disrupted, a phenomenon known as myocardial disarray[1].

There is a significant overlap between the presenting features of both the diseases and diagnosis can be tricky at times. Both give rise to almost same murmur, except that the one caused by AS will be exaggerated by squatting and valsalving, standing suddenly will exaggerate the one caused by HOCM. Diagnosis is mainly by transthoracic 2 D echo or angiography, later one being rarely used now days. The differentiating features of the two diseases are given in Table-1.

On 2 D echo, AS will present as typical stenotic narrowed valve. However, one need to carefully examine the valve to ensure that this indeed is the cause of increased turbulence, specially so if the valve appears to be normal physically. In obstructive variety of HOCM, the obstruction was thought to develop due to systolic anterior motion (SAM) of the mitral valve. Recent echocardiographic evidence however indicates that drag, the pushing force of flow is the dominant hydrodynamic force on the mitral leaflets[2-4]. In obstructive HCM the mitral leaflets are often large and are anteriorly positioned in the LV cavity due to anteriorly positioned papillary muscles that at surgery are often "agglutinated" on to the LV anterior wall by abnormal attachments. The mid septal bulge aggravates the malposition of the valve and re directs outflow so that it comes from a lateral and posterior direction[4]. The abnormally directed outflow may be visualized behind and lateral to the enlarged mitral valve, where it catches it, and pushes it in to the septum[2-4]. There is a crucial overlap between the inflow and outflow portions of the left ventricle[5]. As SAM progresses in early systole the angle between outflow and the protruding mitral leaflet increases. A greater surface area of the leaflets is now exposed to drag which amplifies the force on the leaflets- drag increases with

Table-1: Comparison between HOCM and aortic stenosis on echocardio graphy.

Echocardiography		
	Aortic stenosis	Hocm
Aortic valve calcification	Common	No
Dilated ascending aorta	Common	Rare
Ventricular hypertrophy	Concentric	Asymmetric often involving septum
Physical exam		
Murmur of a1	Common	No
Pulse pressure after pvc	Increased	Decreased
Pulse pressure after valsalva	Decreased intensity	Increased intensity
Carotid pulsation	Normal or tardus et parvus	Brisk, jerky or bisferiens pulse

increasing angle relative to flow[4]. Same thing might have happened in our patient as well. Post op echo in fig-1 suggests that anterior leaflet is perhaps longer than normal and abuts on the septum causing increased gradients. It is easy to confuse the two conditions but an alert echo cardiography operator may be able to pick the right diagnosis and save the patient all the pain.

CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

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