Concomitant Aortoplasty with Septal Myectomy for Supravalvar Aortic Stenosis in an Adult

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ABSTRACT

Survival of patients with severe congenital aortic stenosis beyond third decade is uncommon. This report describes a 31 year old patient who underwent successful aortoplasty and in addition septal myectomy for complete relief of left ventricular outflow obstruction.

Keywords: Aortoplasty; Myectomy; Supravalvar Aortic Stenosis

1. Introduction

Congenital Supravalvar Aortic Stenosis is a rare form of obstructive lesion of the left ventricular outflow tract. The defining feature of the malformation is an aortic narrowing at the level of the sino tubular junction. The commonly associated lesions include peripheral pulmonary artery lesions, coronary lesions, abnormalities of the aortic leaflets and diffuse narrowing of ascending aorta. Survival beyond three decades is uncommon and this report describes a case of supravalvar aortic stenosis in a 31 year old patient who underwent successful aortoplasty and septal myectomy.

2. Case Summary

A 31 years old lady with symptoms of class III dyspnea, intermittent chest pain and syncopal attacks of 2 years duration was referred to our institute. Two Dimensional Echocardiography revealed discrete hour glass type of Supravalvular Aortic Stenosis with severe left ventricular hypertrophy (Figure 1). Left ventricle to aorta peak gradient was 163 mm Hg with a mean of 101 mm Hg. She underwent surgical repair using cardiopulmonary bypass and blood cardioplegic arrest.

Inverted Y shaped Aortotomy was done with limbs extending into non and right coronary sinus. The aortic valve was tricuspid and competent Intraluminally the posterior ridge above left coronary sinus was excised. There was severe obstructive hypertrophy of septum and hence septal myectomy was done. The aortotomy was closed with inverted Y shaped pericardial patch extending into the non coronary sinus and right coronary sinus. The post operative recovery was uneventful. Pre discharge echocardiography (Figure 2(a)) and CT angiographic study (Figure 2(b)) showed a wide open LVOT with a peak gradient of 20 mm Hg and mean of 12 mm Hg.

Ethics Committee approval and patient consent for publication of this data was obtained.

3. Discussion

Congenital supravalvular aortic stenosis often presents in
Childhood and if not corrected by surgery can lead to heart failure and death. The defining feature is a focal or diffuse narrowing starting at the Sino-tubular junction and often involving the entire ascending aorta with rare involvement of the aortic arch and occasionally the peripheral arterial system [1]. This disorder occurs in an autosomal inherited form and in a rare sporadic form it is frequently associated with Williams-Beuren syndrome [2]. The underlying cause has been identified as a mutation of the elastin gene on chromosome 7q11.23 [3]. The first report was in 1878 by Archer who described an elastic band stretching across distal to the aortic cusps [4]. First successful surgical repair with standard patch was described by McGo in 1961 [5]. Doty et al described extended aortoplasty with bifurcated patch in 1977 which allows more symmetric enlargement of the aortic root [6]. Brom described enlarging all the 3 sinuses with ascending aorta enlargement by an additional patch [7]. Surgical treatment of supravalvar aortic stenosis by augmenting the aortic root in 2 or 3 sinuses of Valsalva is associated with a lower mortality rate and fewer reoperations as it preserves the anatomy of the aortic root with better physiologic flow. Currently, septal myectomy is recommended in patients undergoing aortic valve replacement for aortic stenosis with severe basal septal hypertrophy, as it leads to better regression of left ventricular hypertrophy due to more effective reduction of left ventricular outflow tract gradient [8]. The prognosis of supravalvular aortic Stenosis is poor and survival to adulthood without intervention is unusual. The outflow obstruction can be relieved by patch aortoplasty and septal myectomy with good results.

REFERENCES


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