

Subaortic membrane with patent ductus arteriosus

Nishes Basnet, Alka Singh, Rajat Pradhan, Samir Azam Sunny and Asit Baran Adhikary

Article Info

Department of Cardiac Surgery, Faculty of Surgery, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka, Bangladesh

For Correspondence:

Nishes Basnet
nishesbasnet@gmail.com

Received: 23 September 2017
Accepted: 18 November 2017
Available Online: 26 November 2017

ISSN: 2224-7750 (Online)
2074-2908 (Print)

DOI: 10.3329/bsmmuj.v10i4.34646

Cite this article:

Basnet N, Singh A, Pradhan R, Sunny SA, Adhikary AB. Subaortic membrane with patent ductus arteriosus. *Bangabandhu Sheikh Mujib Med Univ J.* 2017; 10: 243-45.

Copyright:

The copyright of this article is retained by the author(s) [Attribution CC-BY 4.0]

Available at:

www.banglajol.info

A Journal of Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

Abstract

A 10 years old boy diagnosed as subaortic membrane with patent ductus arteriosus presented with history of occasional fever, repeated respiratory tract infection since childhood and poor gain weight.

Introduction

Sub aortic membrane is an infrequent disease that produces variable degree of obstruction to the degrees of blood across left ventricular outflow tract.¹ Its prevalence is 1-2% of all congenital cardiac defects and causes 15-20% of all fixed left ventricular outflow tract obstructions.² Disease has overall male preponderance with gender ratio of 2:1 to 3:1.² The prevalence of discrete subaortic stenosis is increasing in adults due to greater number of repaired congenital heart diseases.³ Ventricular septal defect, atrial septal defect, patent ductus arteriosus, coarctation of aorta, bicuspid aortic valve, parachute mitral valve, and persistent left superior venacava are among the associated congenital cardiac defects in 25-50% of the cases. Disease etiology is not completely understood. Various anatomical characteristics that promote chronic flow disturbances in left ventricular outflow tract include a steep aorto septal angle of more than 1300, increased mitral and aortic annular separation, exaggerated override of aorta with misalignment of inter-ventricular septum, sub-aortic shelf and surgical intervention for obstructive lesions during early childhood.^{4,5}

Patent ductus arteriosus is usually found as an isolated lesion in the patients with non-cyanotic congenital heart disease. Association of patent ductus arteriosus with sub aortic membrane is found in about 14% of the patients.⁶ Children usually become symptomatic at the age of 10-20 years. Exertional dyspnea, effort angina and syncope are most common symptoms. On cardiovascular examination, a low pitched ejection systolic murmur in second and third left intercostal space with

radiation to suprasternal notch and absent ejection click is the most important clue for the diagnosis of subaortic membrane. ECG may show left ventricular hypertrophy (50-85%) and strain pattern in 25% of the patients.

Echocardiography is the main imaging study. 2D echo allows definition of exact type, location, extent of left ventricular outflow tract and aortic valve involvement. CW Doppler records peak and mean pressure gradient across left ventricular outflow tract. Trans-esophageal echocardiography allows per operative evaluation of lesion, guides surgical resection and provides immediate surgical results.⁷

Case Report

A 10 year old boy was admitted in the Department of Cardiac Surgery, Bangabandhu Sheikh Mujib Medical University with the history of occasional fever, repeated respiratory tract infection since childhood and poor weight gain. On clinical evaluation there was systolic murmur best heard in the aortic area. Chest X-ray was normal. ECG revealed sinus tachycardia. Echocardiography revealed: Subaortic membrane causing severe aortic valve stenosis (peak pressure gradient was 107 mm Hg). Mitral regurgitation was in Grade II. There was mild tricuspid regurgitation with mild pulmonary hypertension (pulmonary artery systolic pressure: 38 mm Hg). There was a large patent ductus arteriosus (size: 5.8 mm) with left to right shunt with peak pressure gradient of 120 mm Hg.

He was scheduled for patent ductus arteriosus ligation and subaortic membrane resection on 18/3/2017. The standard median sternotomy



was done following general anesthesia.

Pericadiotomy and aorto bi-caval cannulation were done and cardiopulmonary bypass was established. Hypothermia was done at 32°C. Heart was arrested by antegrade blood cardioplegia and the aortotomy was done. Patent ductus arteriosus was identified and ligated by the silk. Subaortic membrane was excised and adequate aortic valve opening was ensured. The aorta was closed by layers. The deairation of the left ventricle was done and the temperature was raised. Patient weaned off cardiopulmonary bypass with minimal inotropic support. He had an uneventful post-operative recovery.

Discussion

In a study of 35 patients, the prevalence of associated other congenital cardiac defects was 65% [patent ductus arteriosus (34%), VSD (20%), coarctation (23%), PS (9%) and miscellaneous defects (9%).⁸ In our case, it was found to be associated with patent ductus arteriosus. Association of subaortic stenosis with tetralogy of fallot, especially in women, is extremely rare. The etiology of discrete subaortic stenosis in general and in association with patent ductus arteriosus is unclear.⁹ This endocardial abnormality involves the subaortic ridge and the leaflets of the adjacent valves.¹⁰ Those patients who have left ventricle-aorta gradients beyond 30 mm Hg¹¹ or has a co-existing cardiac defect requires surgical correction while some authors advocate surgical resection for subaortic membrane for any degree of obstruction because aortic insufficiency, left ventricular hypertrophy and infective endocarditis may develop due to subaortic stenosis Surgical.^{12,13} As in our case the patient had patent ductus arteriosus with peak pressure gradient 107 mm Hg. It is also said that recurrence and reoperation can be prevented by surgical resection of fixed subaortic stenosis before the development of a significant outflow tract gradient i.e., >40 mm Hg.¹⁴

The most favorable surgical method for patients with subaortic membrane is questionable. Enucleation of the discrete membrane with its fibromuscular ridge is the most preferable method for some surgeons, while other surgeons do the resection of the hypertrophied muscle with routine myomectomy. The occurrence of late aortic regurgitation can be reduced by radical excision of all diseased tissue.¹⁵ But this aggressive process may increase the risk of iatrogenic damage to the conduction tissue (injury to the conduction tissue between the right and noncoronary cusps), mitral valve and ventricular septal defect. In this case, ligation of the patent ductus arteriosus was done then radical excision of the diseased tissue was

done and adequate opening of the aortic valve was secured.

There is significant rate of recurrence of subaortic stenosis but cause of recurrence for left ventricular outflow tract obstruction is still unknown even after adequate excision. Routine removal of underlying septal muscle may prevent recurrence which may be an important cause as it is the initial site of fibromuscular obstruction.¹⁶

References

1. Iwata Y, Imai Y, Shin'oka T, Kurosawa H. Subaortic stenosis associated with systolic anterior motion. *Heart Vessels*. 2008; 23: 436-39.
2. Hoffman JI, Christianson R. Congenital heart disease in a cohort of 19,502 births with long-term follow-up. *Am J Cardiol*. 1978; 42: 641-47.
3. Oliver JM, González A, Gallego P, Sánchez-Recalde A, Benito F, Mesa JM. Discrete subaortic stenosis in adults: Increased prevalence and slow rate of progression of the obstruction and aortic regurgitation. *J Am Coll Cardiol*. 2001; 38: 835-42.
4. Bezold LI, Smith EO, Kelly K, Colan SD, Gauvreau K, Geva T. Development and validation of an echocardiographic model for predicting progression of discrete subaortic stenosis in children. *Am J Cardiol*. 1998; 81: 314-20.
5. Rohlicek C, del Pino SF, Hosking M, Miro J, Cote JM, Finley J. Natural history and surgical outcomes for isolated discrete subaortic stenosis in children. *Heart* 1999; 82: 708-13.
6. Laurel S, Kathryn HE, Aaron RL, Mary AE. Membranous sub aortic stenosis and patent ductus arteriosus. *Chest J*. 1977; 72: 333-38.
7. Barkhordarian R, Wen-Hong D, Li W, Josen M, Henein M, Ho SY. Geometry of the left ventricular outflow tract in fixed subaortic stenosis and intact ventricular septum: an echocardiographic study in children and adults. *J Thorac Cardiovasc Surg*. 2007; 133: 196-203.
8. Motro M, Schneeweiss A, Shem-Tov A, Benjamin P, Kaplinsky E, Hegesh J. Correlation of distance from subaortic membrane to base of the right aortic valve cusp and the development of aortic regurgitation in mild discrete subaortic stenosis. *Am J Cardiol*. 1989; 64: 395-96.
9. Leichter DA, Sullivan I, Gersony WM. Acquired discrete subvalvular aortic stenosis: Natural history and hemodynamics. *J Am Coll Cardiol*. 1989; 14: 1539-44.
10. Troyer JR. Development of the heart and the fetal circulation. In: *Cardiovascular disorders*. Brest AN, Moyer JH (eds). Philadelphia, FA Davis and Co, 1968, p 20.
11. Kirklin JW, Barratt-Boyes BG. Cardiac surgery:

- Morphology, diagnostic criteria, natural history, techniques, results, and indications. 2nd ed. Vol 2. New York, Churchill Livingstone, 1993, pp 1212-24.
12. Shamsuddin AK, Al-Manzoor MIS, Kabiruzzaman M. Isolated Fixed discrete subaortic membrane: A case report. *Bangladesh Heart J.* 2015; 30: 43-45.
 13. Douville EC, Sade RM, Crawford FA Jr, Wiles HB. Subvalvar aortic stenosis: Timing of operation. *Ann Thorac Surg.* 1990; 50: 29-34.
 14. Oliver JM, Gonzalez A, Gallego P, Sanchez-Recalde A, Benito F, Mesa JM. Discrete subaortic stenosis in adults: increased prevalence and slow rate of progression of the obstruction and aortic regurgitation. *J Am Coll Cardiol.* 2001; 38: 835-42.
 15. Ronbrauner H, Laks F, Davis C, Drinkwater. Benefits of early surgical repair in fixed subaortic stenosis. *J Am Coll Cardiol.* 1997; 30: 1835-42.
 16. Parry AJ, Kovalchin JP, Suda K, McElhinney DB, Wudel J, Silverman NH, et al. Resection of subaortic stenosis; can a more aggressive approach be justified? *Eur J Cardiothorac Surg.* 1999; 15: 631-38.
 17. Lupinetti FM, Pridjian AK, Callow RN, Crowley DC, Beekman RH, Bove EL. Optimum treatment of discrete subaortic stenosis. *Ann Thorac Surg.* 1992; 54: 467-71.
-