Subaortic membrane with patent ductus arteriosus

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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. Its prevalence is 1-2% of all congenital cardiac defects and causes 15-20% of all fixed left ventricular outflow tract obstructions. 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. Ventricul 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 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 aortic 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.

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. 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. Transesophageal echocardiography allows per operative evaluation of lesion, guides surgical resection and provides immediate surgical results.

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 murmurm 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 32ºC. 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 deaira-
ture was raised. Patient weaned off cardiopulmo-
nary 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%), coarcta-
tion (23%), PS (9%) and miscellaneous defects (9)]. In
our case, it was found to be associated with
patent ductus arteriosus. Association of subaortic
stenosis with tetrology 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. This endo-
cardial abnormality involves the subaortic ridge and
the leaflets of the adjacent valves. Those patients
who have left ventricle-aorta gradients beyond 30 mm Hg 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.

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.

The most favorable surgical method for patients
with subaortic membrane is questionable. Enuclea-
tion of the discrete membrane with its fibro-
muscular 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. 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 sepal 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.

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