Case Report
Pak Armed Forces Med J 2015; 65(Suppl): S155-57

SITUS INVERSUS, BICUSPID AORTIC VALVE, SUBAORTIC MEMBRANE AND AORTIC STENOSIS-A RARE COMBINATION

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ABSTRACT

Situs inversus, dextrocardia with subarctic membrane and bicuspid aortic valve is a rare entity, making surgical procedures complicated and challenging. We describe a case of a 23 year old male patient, presenting with a history of dyspnoea on exertion. Lung fields were clear and the apical impulse was palpable in the fifth inter costal space in the right mid-clavicular line. A chest roentogram showed dextrocardia, an elevated /t hemi-diaphragm with the gastric bubble on the right side Aortic valve stenosis with dextrocardia was suspected from the history and clinical examination and a transthoracic echocardiogram (TTE) was preformed. Surgical setup was the same as for routine cardiac surgical procedures; with the exception that the surgeon was on the opposite side (left side of the patient). The postoperative course was uncomplicated and the patient remains asymptomatic at one year follow up. His recent echos show regression in ventricular hypertrophy and acceptable gradients across the prosthetic aortic valve.

Keywords: Adult congenital heart diseases, Situs inversus, Bicuspid aortic valve, Subaortic membrane.

INTRODUCTION

Operating on patients with dextrocardia presents special challenges during cardiac surgery as the entire operating field needs to be modified for the surgery. We present an adult case, with situs inversus, dextrocardia, bicuspid aortic valve with aortic stenosis and subaortic membrane, a so far unreported association.

CASE REPORT

A 23 year old male patient, presented with a history of dyspnoea on exertion. He had no history of diabetes, hypertension or hyperlipidemia. He denied any history of chest pain, syncope, fever, respiratory infections or illicit drug use. He had percutaneous ballon valvuloplasty of the aortic valve at the age of 8 years. There was no family history of congenital heart disease.

Physical examination showed a slow rising pulse with a normal sinus rhythm. Lung fields were clear and the apical impulse was palpable in the fifth intercostal space in the right mid clavicular line. There was a palpable systolic thrill over the right precordium. On auscultation there was normal intensity S1 and S2. A grade IV ejection systolic murmur was audible over the upper right sternal border with radiation to the carotids. All peripheral pulses were palpable.

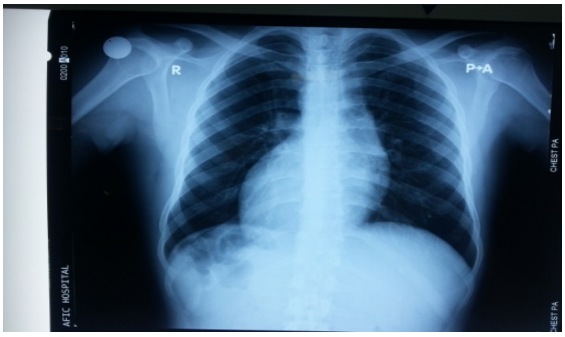

Figure-1: Situs inversus, Dextrocardia prominent Rt heart border due to left ventricular hypertrophy.

A chest roentogram showed dextrocardia, an elevated /t hemidiaphragm with the gastric bubble on the right side (Fig-1). Aortic valve stenosis with dextrocardia was suspected from the history and clinical examination and a transthoracic echocardiogram (TTE) was preformed. TTE revealed normal left ventricular function with gross left ventricular hypertrophy, and a septal thickness of 19-23 mm. There was severe aortic stenosis, with a bicuspid aortic valve. The PPG across the LVOT / Aortic valve was 100 mmHg. There was a discrete sub-aortic fibro muscular membrane. Aortic valve replacement with resection of the subaortic membrane was planned. Preoperative

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workup was done and the patient prepared for surgery.

Surgical setup was the same as for routine cardiac surgical procedures; with the exception that the surgeon was on the opposite side (left side of the patient). Cardiac exposure included a midline sternotomy and a pericardotomy. Cardiopulmonary bypass was achieved with aortic and bicaval cannulation. Cardiac arrest was attained through antegrade cold blood cardioplegia. An oblique aortotomy was done. The diseased stenotic bicuspid aortic valve was excised (Fig-2). The fibromuscular subaortic membrane was identified and excised; limited myomectomy of the left ventricular outflow trackto permita size 24 hegar dialator was done. A 19 mm St Jude HP mechanical valve was secured at the aortic position (Fig-3). We closed the aorta in two layers.

The postoperative course was uncomplicated and the patient remains asymptomatic at one year follow up. His recent echos show regression in ventricular hypertrophy and acceptable gradients across the prosthetic aortic valve.

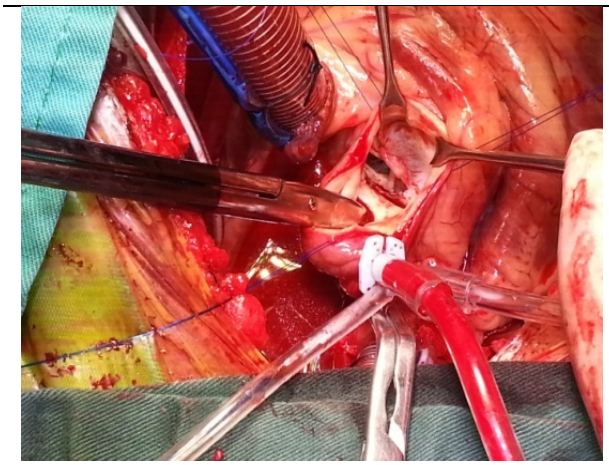

Figure-2: Aortotomy with the scissor tip pointing at the left main coronary ostium. Bicuspid aortic valve excised. Fibromuscular sub aortic membrane visible beneath the aortic annulus.

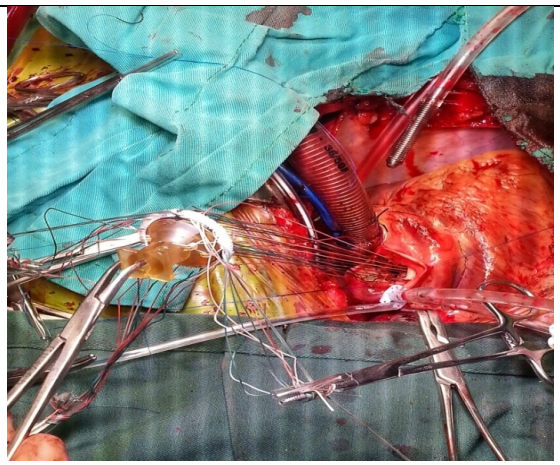

Figure-3: Implanting a mechanical bileaflet mechanical valve at aortic position.

DISCUSSION

Marco Severino described dextrocardia in 1643. More than a century later, Matthew Baillie described the complete mirror-image reversal of the thoracic and abdominal viscera known as situs inversus. Situs inversus is present in 1/10,000 of the population[1]. The incidence of congenital heart disease with situs inversus is 3-5%. Situs inversus with levocardia is rare and is invariably always associated with congenital heart disease[2].

Bicuspid aortic valve is present in 1-2% of the population. Patients with bicuspid aortic valve may be completely asymptomatic, About 30-40% of these patients present with complications. Symptoms when present result from the valve causing stenosis or insufficiency.

Aortic stenosis is a common cardiac problem. Aortic stenosis can be due to valvular, subvalvular and supravalvular causes[3]. Subvalvular causes include subaortic rings and membranes, which present at a young age, depending on the size of the ring and the degree of obstruction. The common causes of aortic stenosis are rheumatic heart disease, degenerative calcific aortic stenosis and bicuspid aortic valve. Patients usually present with symptoms of angina, syncopal episodes and heart failure. Symptoms often develop after a latent period of 10-20 years. Once symptomatic these patients have 25% mortality at 1 year which rises to about 50% at 2 years. Aortic valve replacement improves symptoms

and prognosis and is the treatment of choice for severe aortic stenosis. Percutaneous balloon valvuloplasty/TAVI is a palliative choice in critically ill patient who are not surgical candidates.

Subaortic stenosis (SAS) is a wide spectrum of anatomic anomalies ranging from a discrete fibrous membrane to a narrow tubular tunnel, causing obstruction to the left ventricular outflow tract. This is an uncommon cause accounting for 8-30% of Lt sided obstructive lesions. SAS often coexists with other congenital anomalies in 45-62% of the cases[5]. The majority of these patients have an abnormal aortic valve. Due to the progressive nature of SAS and associated complications, early surgery is indicated. The ideal treatment for subaortic stenosis involves excision of the membrane along with septal myomectomy[4]. An aggressive surgical approach is advocated in the management of SAS as this is associated with better long term results and decreased incidence of restenosis[5].

Cardiac surgery in dextrocardia is technically difficult due to anatomic malpositioning. Modifications are required in the setup and surgical technique to facilitate exposure and surgery[6].

Conflict of Interest

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

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