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Original Research Paper

Cardiology

LARGE ATRIO-VENTRICULAR CANAL DEFECT WITH BIDIRECTIONAL SHUNT WITH SEVERE PULMONARY STENOSIS WITH SPUTUM POSITIVE PULMONARY KOCH'S – A CASE REPORT.

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ABSTRACT Congenital heart defects (CHD) are cardiovascular malformations that generally occur due to aberrant development of a normal structure in the fetus, or failure of such a structure to progress beyond an early stage of embryonic or fetal development.
Malformations are due to complex multi-factorial genetic and environmental causes. Congenital heart defects, in a definition proposed by Mitchell et al, is a gross structural abnormality of the heart or intrathoracic great vessels that are actually or potentially of functional significance. A comprehensive approach to every aspect of CHD, covering from
embryology, fetal malformations, pathology, clinical approach, investigations, interventions to the surgery is very essential to reduce the morbidity and mortality in children with CHD. Here is a case of young male with complex congenital heart defects with marfanoid habitus and diabetes mellitus with sputum positive pulmonary koch's.

KEYWORDS : Congenital malformation, Great vessels, Fetus, Pulmonary koch's.

INTRODUCTION

Congenital heart defects (CHD) are the commonest of all congenital Lesions and the most common type of heart defects among children[1].These defects generally result from the aberrant development of a normal structure in fetus or failure of progress beyond the early stage of embryonic or early fetal development[2]. Cardiac mal-development early in the embryo leads to significant morbidity and mortality. The CHD as defined by Mitchell et al is a gross structural abnormality of the heart or intra thoracic great vessels that are actually significance[3]. The estimated prevalence of CHD is 8 to 10 per 1000 live births, with a higher rate of stillbirth, spontaneous abortion, and prematurity[4]. The relative frequency of the most common lesions varies with different reports but nine common lesions form 80% of congenital heart defects[5].Recent advances in diagnosis and surgical treatment over the past 40 years have led to dramatic increases in survival for children with serious heart defects.

CASE REPORT :

A 27 year old male with history of cyanotic spells in his childhood , presented to our hospital with low grade continuous fever and progressive dyspnea of one month duration. Newly detected diabetes mellitus. Cardiac examination revealed grade 3 parasternal heave with loud P2 component of second heart sound. Respiratory system examination revealed bilateral infraxillary fine crackles. He too had marfanoid habitus in the form of long stature, arm span more than height, high arched palate with positive thumb and wrist sign. Patient was evaluated for congenital heart disease and found to have a large atrio-ventricular canal defect with bidirectional shunt with large ostium primum atrial septal defect with large ventricular septal defect with severe pulmonary stenosis on 2 D Echocardiogram. Sputum was positive for acid fast bacilli. Further, was started with Anti-tubercular treatment and blood glucose levels were controlled with insulin and he is better on follow up.

DISCUSSION

The etiological factor of most Congenital heart defects is unknown. In the present scenario, the complex genetics, and inheritance of CHD remains incompletely understood. In the past, the circumstances were even more worst because many children with CHD did not survive to reproductive age and fetal echocar diography was not available. In most of the cases, it is multi-factorial in origin and is a result of both genetic predisposition and environmental factors. Known genetic causes of heart disease includes inherited chromosomal abnormalities such as trisomy 21, 13, and 18, as well as a range of newly recognized genetic point mutations, point deletions and other genetic abnormalities as seen in syndromes such as CATCH 22, familial ASD with heart block, Alagille syndrome, Noonan syndrome, and many more. Complete atrioventricular canal (CAVC), also referred to as complete atrioventricular septal defect, is characterised by an ostium primum atrial septal defect, a common atrioventricular valve and a variable deficiency of the ventricular septum inflow. CAVC is an uncommon congenital heart disease, accounting for about 3% of cardiac malformations. Atrioventricular canal occurs in two out of every 10,000 live births. Both sexes are equally affected and a striking association with Down syndrome was found. Depending on the morphology of the superior leaflet of the common atrioventricular valve, 3 types of CAVC have been delineated (type A, B and C, according to Rastelli's classification). CAVC results in a significant interatrial and interventricular systemic-to pulmonary shunt, thus inducing right ventricular pressure and volume overload and pulmonary hypertension[6]. Diagnosis of CAVC might be clinically suspected in patients presenting in the first few months of life with congestive heart failure, cardiomegaly on chest X-ray and left axis deviation, bi-atrial enlargement and bi-ventricular pressure and volume overload on electrocardiogram (ECG). Echocardiography is the key tool for the diagnosis and anatomic classification of this malformation. It shows the ostium primum atrial septal defect, with the underlying common atrioventricular valve, and the defect of the ventricular septal inflow.

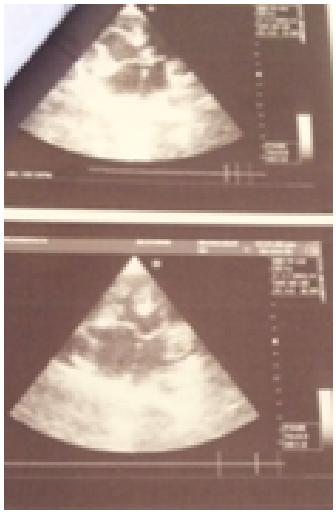

FIG 1 : 2 D ECHO

FIG 2 : WRIST SIGN

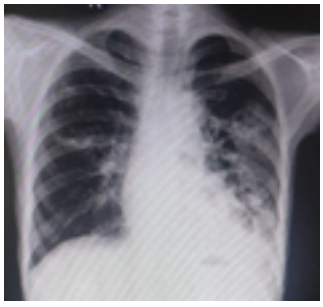

FIG 3 : CHEST RADIOGRAPH

CONCLUSION :

Large atrio-ventricular (AV) canal defect with bidirectional shunt with large ostium primum atrial septal defect (ASD) is a rarest case among congenital heart disease. In addition the patient also had sputum positive pulmonary koch's , may be secondary to immune compromised state as a result of diabetes mellitus and chronic cardiac illness. So it is of utmost importance to detect these kind of complex congential cardiac conditions in early phase of life for necessary correction , If failure to do so then the patient may have an inoperable state in the later adulthood with highest amount of morbidity and mortality.

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