

An isolated cleft of the anterior leaflet of mitral valve in an infant: a case report

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ABSTRACT

Background: Isolated cleft of the anterior leaflet of the mitral valve is the occurrence of mitral cleft without ostium primum or ostium secundum defect and it is rare. It results from different degrees of failure of fusion of early embryonic atrioventricular endocardial cushions and occurs more in children with trisomy 21. It commonly presents with mitral incompetence which worsens as the child grows.

Aim: To document a case of congenital isolated cleft of the anterior leaflet of the mitral valve in an infant. **Findings:** A nine month old boy presented in our clinic with a two month history of fast breathing, cough, and loss of weight. He was found not to be cyanosed but pale, tachypnoeic, dyspnoeic with basal crepitations. Cardiovascular system examination revealed he had a precordial bulge with tachycardia, cardiomegaly and a holosystolic murmur maximum at the apex. There was tender hepatomegaly. He was diagnosed as having congestive cardiac failure with broncho-pneumonia secondary to an acyanotic congenital heart disease. Plain chest X-ray confirmed the cardiomegaly and bronchopneumonia while an electrocardiography showed bilateral atrial enlargement with right ventricular hypertrophy. The cardiac failure was treated but he represented with recurrent heart failure upto three times within six months. An echocardiogram done on the third admission showed an isolated cleft of the anterior leaflet of the mitral valve and he has been referred for surgery.

Conclusion: Isolated cleft of the mitral valve commonly presents with mitral incompetence and eventual heart failure as in our patient. Early surgical intervention is advised as the width of the cleft tend to increase as the child grows leading to worsening of the mitral insufficiency. The definitive treatment is direct closure of the cleft. The presentation also highlights the challenges of medical practice in resource-poor countries.

Key words: Congenital , heart, isolated cleft, mitral valve, ostium primum, ostium secundum

INTRODUCTION

Isolated cleft of the anterior leaflet of the mitral valve is the occurrence of mitral cleft without ostium primum or ostium secundum defect.^[1] It is a rare occurrence and a rare cause of mitral insufficiency.^[2] Clinically significant congenital mitral valve lesions are rare and estimated to affect 0.4% of those with congenital heart disease or 5/100,000 of the general population.^[3] Usually, mitral cleft with or without ostium primum defect is associated with other congenital heart defects including ventricular septal defect, tetralogy of Fallot, tricuspid atresia, double-inlet left ventricle.^[4] Its occurrence with an anomalous origin of the left coronary artery leads to death in over 90% of patients in the first year of life without treatment.^[5] Congenital cleft of the mitral valve is a rare cause of mitral incompetence, resulting from various degrees of failure of fusion of the embryonic atrioventricular (AV) endocardial cushions.^[6] Sigfusson and colleagues suggested that a cleft in an otherwise normal mitral valve should be classified separately from atrioventricular canal defects (AVCD) with a common junction.^[7] In isolated cleft of the anterior mitral leaflet, the annulus is in the normal position and incompetence is caused by flail segments of the anterior leaflet.^[8] The most common clinical presentation is congestive cardiac failure secondary to mitral incompetence.^[9] We present this infant boy who presented to us with congestive heart failure whom we diagnosed to have acyanotic congenital heart disease queried ventricular septal defect but which came out to be anterior cleft of the leaflet of the mitral. The presentation and literature review of isolated cleft of the anterior leaflet of the mitral valve is presented. To the best of our knowledge, this is the first documentation in Nigeria.

CASE REPORT

The patient was first seen at our clinic at the age of nine months on 12/12/2011 with a three month history of cough, fast breathing, diarrhoea and was found to be dyspnoeic, tachypnoeic, pale and with diaphoresis. There was no cyanosis, but he was underweight with a weight of 7kg (expected is 9kg). He had tachycardia with a heart rate of 150beats/minute. He had a precordial bulge with apex beat at the 5th left intercostal space mid-clavicular line.

The first and second heart sounds were heard and normal but he had a grade 3/6 holosystolic murmur maximum at the apex and radiating to the back. The respiratory rate was 84cycles/minute with flaring of the alar nasi, intercostal and subcostal recessions. He also had crepitations at the lower zone of the right lung anteriorly. There was tender hepatomegaly on abdominal palpation. A diagnosis of acyanotic congenital heart disease with congestive cardiac failure with bronchopneumonia was made and the following investigations ordered for: full blood count, plain chest X-ray, electrocardiography and echocardiography.

RESULTS

Table 1: Haematological indices and blood group

Item	Results (Normal ranges in brackets)
Haemoglobin	9.3g/dl (14-18g/dl)
PCV	28.9%(40-54%)
MCV	78.5(80-95fl)
MCH	25.2(27-32pg)
MCHC	32.1(30-35Ppg/dl)
Total white cells	34,900 (4000-11,000cells/mm)
Granulocytes	85.4%
Lymphocytes	9.4%
Monocytes	5.2%
Platelets count	529(150-400)
Blood group	B+

The results show anaemia with leucocytosis, and absolute granulocytosis with thrombocytosis. The plain chest X-ray (CXR) showed cardiomegaly with patchy opacities both lung fields. The electrocardiography (ECG) showed a sinus rhythm with normal axis considering the age, bilateral atrial enlargement with right ventricular hypertrophy with repolarization abnormality.

He was treated with frusemide then later with spironolactone and hydrochlorothiazide, and ceftriazone which was later changed to cefixime suspension and was discharged after two months on admission. However, he was re-admitted just one week after discharged with

congestive cardiac failure (CCF) and the weight had dropped to 6.5kg. He stayed for a week and was discharged due to parental pressure. He was re-admitted for the third time for CCF in April 2012, four months from the initial admission. The echocardiogram showed he had cleft of the anterior leaflet of the mitral valve. He was treated and referred to India for surgery after two weeks of admission. While waiting and arranging for the logistics, he represented for the fourth time with CCF two months later and was last seen on 27th June 2012, that is a week after discharge for follow up.

DISCUSSION AND LITERATURE REVIEW

Cleft, derived from the verb to *cleave*, is defined as a space or opening made by splitting.^[10] A cleft mitral valve has a split anterior with each part of the leaflet typically attaching to a different papillary muscle.^[10] Our patient presented early in life, in infancy. Just as in our patient, it commonly presents with congestive heart failure secondary to mitral incompetence. Early presentation in our patient may be as a result of associated other cardiac anomalies which were not detected with the 2-D echocardiography. It has been documented that there could be discordance between the echocardiographic and surgical/postmortem findings in mitral cleft.^[11] Congenital cleft malformation in an otherwise normal mitral valve usually presents with concomitant cardiac defects, mainly an atrial septal defect, and Down's syndrome is the commonest common noncardiac anomaly.^[12] It is known that its association with anomalous origin of the left coronary artery presents early in infancy with congestive cardiac heart failure and death occurs in infancy in over 90% of cases if surgery is not done in the isolated cleft. The anterior mitral leaflet is best visualized from a subcostal or a parasternal axis view.^[13] From that parasternal long axis view, the presence of the cleft could be suspected based on an abnormal orientation of the anterior mitral leaflet towards the outflow septum.^[13] When available, colour doppler mapping clearly demonstrates the location and extent of mitral regurgitation.^[14] Also, colour and septal doppler identified left ventricular outflow obstruction caused by the mitral cleft attachments.^[14] The width of the cleft in some cases with normally

related great arteries appear to increase with age and because the mitral regurgitation in isolated cleft of the mitral valve is usually progressive, early surgical intervention is recommended even when the mitral regurgitation is mild.^[4,15] Our patient presented with more frequently recurrence of congestive cardiac failure. Direct suture of the cleft is the preferred procedure, but glutaraldehyde-treated autologous pericardium can be used if there is lack of valvular tissue.^[16] Mitral valve replacement is performed in adult patients whose valves cannot be repaired initially.^[17] The most important complication of cleft repair is need for reoperation.^[4,18] Complete correction of the mitral valve insufficiency is the most important factor affecting long-term complication.^[17,18] A rare long term complication which occurs when the regurgitation is not completely corrected which can occur if the surgeon fears mitral stenosis, is continued mitral regurgitation with accompanying marked left ventricular hypertrophy and dysfunction. This will then require cardiac transplantation.^[4,19]

CONCLUSION

This child would have benefited from early surgical intervention, but because of lack of facilities and poor financial power, the child suffered recurrent congestive cardiac failure.

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