

Case Report

Isolated cleft of the anteroir leaflet of the mitral valve: Case report in a nine month old boy at Delta State University Teaching Hospital, Oghara, Niger Delta, Nigeria

G. I. McGil Ugwu¹ and Umerie EM²

¹Department of Paediatrics, University Teaching Hospital, Oghara, Delta State, Nigeria.

²Department of Medicine, University Teaching Hospital, Oghara, Delta State, Nigeria.

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Isolated cleft of the anterior leaflet of the mitral valve is the occurrence of mitral cleft without ostium primum or ostio secundum defect and it is rare. We present a nine month old boy who 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 showed that 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. 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.

Key words: Cleft, anterior leaflet, mitral valve.

INTRODUCTION

Isolated cleft of the anterior leaflet of the mitral valve is the occurrence of mitral cleft without ostium primum or ostium secundum defect (Mohanty et al., 1999). It is a rare occurrence and a rare cause of mitral insufficiency (Kaan et al., 2003). 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 (Hoffman and Laplan, 2002). Usually, mitral cleft with or without ostium primun defect is associated with other congenital heart defects including

ventricular septal defect, tetralogy of Fallot, tricuspid atresia, and double-inlet left ventricle (Van Praagh et al., 2003). 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 (Rathinam et al., 2005). Congenital cleft of the mitral valve is a rare cause of mitral incompetence, resulting from various degrees of failure of fussion of the embryonic atriventricular (AV) endocardial cushions (McDonald et al., 1994). Sigfussion et al. (1995) suggested that a cleft in an otherwise normal

Table 1. Haematology.

Item	Results (normal ranges in brackets)
Haemoglobin (g/dl)	9.3 (14-18)
PCV (%)	28.9 (40-54)
MCV (fl)	78.5 (80-95)
MCH (pg)	25.2 (27-32)
MCHC (pg/dl)	32.1 (30-35)
Total white cells	34,900 (4000-11000)
Granulocytes (%)	85.4
Lymphocytes (%)	9.4
Monocytes (%)	5.2
Platelets count	529 (150-400)
Blood group	B+ve

PCV: Packed cell volume; MCV: mean cell volume; MCH: mean cell haemoglobin; MCHC: mean cell haemoglobin concentration.

mitral valve should be classified separately from atrioventricular canal defects (AVCD) with a common junction. 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 (Nadas, 1972). The most common clinical presentation is congestive cardiac failure secondary to mitral incompetence (Tamura et al., 2000). We present an infant boy who presented with congestive heart failure whom we diagnosed to have acyanotic congenital heart disease, ventricular septal defect, but which came out to be cleft of the anterior leaflet of the mitral valve. The presentation and literature review of isolated cleft of the anterior leaflet of the mitral valve is presented here, and to the best of our knowledge this is the first documentation in Nigeria.

CASE PRESENTATION

This baby was first seen in our clinic at the age of nine months on 12/12/2011 with 3-month history of cough, fast breathing, diarrhoea. He was found to be dyspnoeic tachypnoeic with diaphoresis and pale. There was no cyanosis, but was underweight with a weight of 7 kg (expected is 9 kg). He had tachycardia with a heart rate of 150 beats/min. 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 84 breaths/min 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 bronch pneumonia was made and the following investigations ordered for: full blood count, plain chest X-ray (CXR), electrocardiography (ECG) and echocardiography.

RESULTS

The results show anaemia with leucocytosis, and absolute granulocytosis with thrombocytosis. The CXR

showed cardiomegaly with patchy opacities in both lung fields. The ECG showed a sinus rhythm with normal axis considering the age, bilateral atrial enlargement with right ventricular hypertrophy with repolarization abnormality (Table 1).

He was treated with frusemide then later with spirinolactone and hydrochlorothiazide, ceftriazone which was later changed to cefixime suspension and was discharged after two months admission. However, he was readmitted just one week after being discharged with congestive cardiac failure and the weight had dropped to 6.5 kg. He stayed for a week and was discharged due to parental pressure. He was readmitted for the third time for congestive heart failure in April 2012, which was 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 congestive cardiac failure two months later and was last seen on 27th June 2012, that is a week after discharge for follow up.

DISCUSSION

Cleft, derived from the verb *to cleave*, is defined as a space or opening made by splitting (Anderson et al., 1985). A cleft mitral valve has a split anterior with each part of the leaflet typically attaching to a different papillary muscle (Anderson et al., 1985). This patient presented early in life, in infancy. Just as in this patient, it commonly presents with congestive heart failure secondary to mitral incompetence. Early presentation in this 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 are at times discordance between the echocardiographic and surgical/postmortem findings in mitral cleft (Smalhorn et al., 1982). Congenital

cleft malformation in an otherwise normal mitral valve usually presents with concomitant cardiac defects, mainly an atrial septal defect, and Down syndrome is the commonest noncardiac anomaly (Fraisie et al., 2002). 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 (McDonald et al., 1994). Cleft in the anterior mitral leaflet is best visualized from a subcostal or a parasternal axis view (Sugeng et al., 2008). 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 (Sugeng et al., 2008). When available, colour Doppler mapping clearly demonstrates the location and extent of mitral regurgitation (Di Segni et al., 1992). Also, colour and septal Doppler identified left ventricular outflow obstruction caused by the mitral cleft attachments (Di Segni et al., 1992). 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 (Van Praagh et al., 2003; Zias et al., 1998). Our patient presented 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 (Stelin et al., 2010). Mitral valve replacement is performed in adult patients whose valves cannot be repaired initially (Perier and Clausnizer, 1995). The most important complication of cleft repair is the need for reoperation (Van Praagh et al., 2003; Ohno et al., 1999). Complete correction of the mitral valve insufficiency is the most important factor affecting long-term complication (Perier and Clausnizer, 1995; Ohno et al., 1999). 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 (Van Praagh et al., 2003; Aharon et al., 1994).

Conclusion

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

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