CASE STUDY

Juvenile Rheumatic Mitral Stenosis with Multiple Ventricular Septal Defects

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ABSTRACT

Congenital heart disease is related to events occurring in the embryonal stage, while rheumatic heart disease is a sequela of immune-mediated damage following streptococcal infection. We report an unusual association of multiple ventricular septal defects and severe pulmonary arterial hypertension with rheumatic mitral stenosis in a 7-year-old girl. This case highlights the need for careful examination for coexisting rheumatic disease in late presentations of congenital heart disease.

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INTRODUCTION

Rheumatic heart disease (RHD) continues to be a significant health problem in India, leading to morbidity and mortality. Rigorous screening has made early diagnosis and timely surgical correction of congenital heart defects possible, but late presentations are occasionally seen, with attendant complications. We discuss an unusual presentation of a child with multiple ventricular septal defects (VSDs) and rheumatic mitral stenosis.

CASE REPORT

A 7-year-old girl was referred for evaluation of worsening exertional breathlessness and palpitations. Two episodes of fever with migratory polyarthritis over the previous 8 months were treated with analgesics by the family physician. There was significant history of poor weight gain during early childhood. The child weighed 12 kg; expected weight for age was 22 kg. Her general physical examination was normal. A hyperkinetic precordium, cardiomegaly with a heaving apex, a loud second heart sound, and a grade 4/6 pansystolic precordial murmur were noted. Chest radiography revealed cardiomegaly with plethora. An electrocardiogram showed sinus rhythm, left axis deviation, and biventricular hypertrophy. Two-dimensional echocardiography demonstrated a thickened stenotic mitral valve (area, 1.1 cm^2) with commissural fusion and thickening of the subvalvar apparatus. Two VSDs were identified, the larger of which was located in the mid-muscular septum, and the smaller in the perimembranous region (Figures 1a and 1b). The patient's clinical condition was stabilized on a regimen of complete bed rest and decongestive therapy. A cardiac catheterization study indicated a left-to-right shunt with severe pulmonary artery hypertension and a high pulmonary vascular resistance index (Table 1). Although definitive evidence of prior streptococcal infection was lacking, a diagnosis of probable rheumatic mitral stenosis with multiple VSDs and severe pulmonary artery hypertension was accepted on the basis of significant history of fever with migratory polyarthritis and the echocardiographic appearance of the valve. A decision was made to operate on the child in view of her symptoms, the presence of significant mitral stenosis, and increased left-to-right shunting with a considerable fall in the pulmonary vascular resistance index on oxygen administration during cardiac catheterization (Table 1). Surgical correction was carried out under standard cardiopulmonary bypass. Transseptal inspection of the mitral valve revealed thickened leaflets, commissural fusion, and thickening of the subvalvar apparatus (Figure 2a). Dacron patch closure of the VSDs, and Rumel debridement of the anterior mitral leaflet, mitral commissurotomy, and commissural plication were carried

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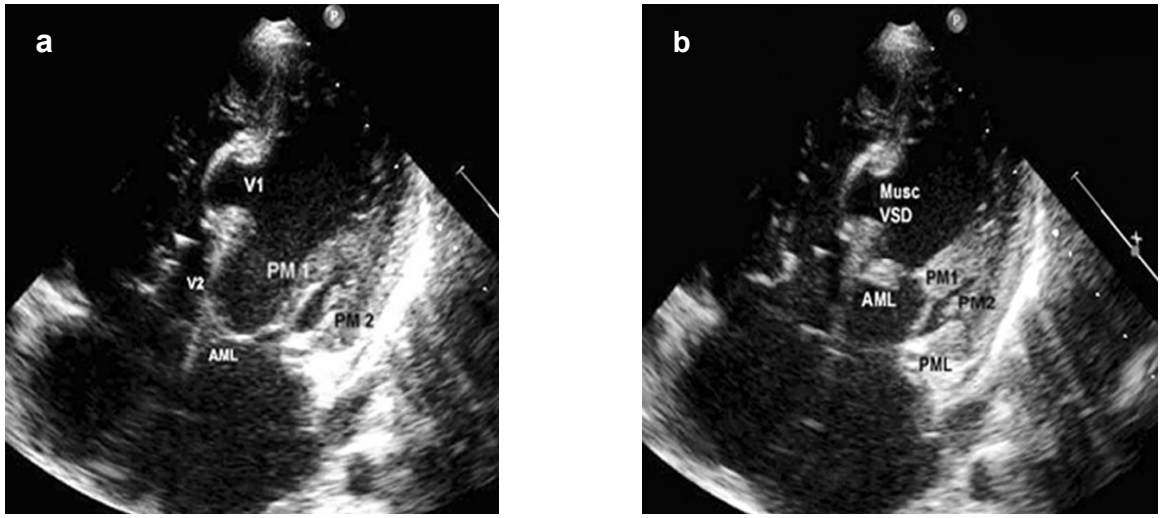

Figure 1. (**a**) Profile of VSD, (**b**) Detail of thickened anterior mitral leaflet and subvalvar apparatus. AML = anterior mitral leaflet, PM1, PM2 = papillary muscles, PML = posterior mitral leaflet, V1 = mid-muscular defect, V2 = perimembranous defect.

Table 1. **Findings of Cardiac Catheterization Study before and 5 min after O_2 Administration**

Parameter	Baseline	100% Oxygen
Mean oxygen step-up (right ventricular level)	20%	24%
Pulmonary artery pressure (mm Hg)	110/60 (74)	95/45 (54)
Pulmonary-systemic shunt ratio (Qp/Qs)	2.5	3.2
Pulmonary vascular resistance index (dyne·s·cm^{-5}·m^{-2})	6.05	2.86

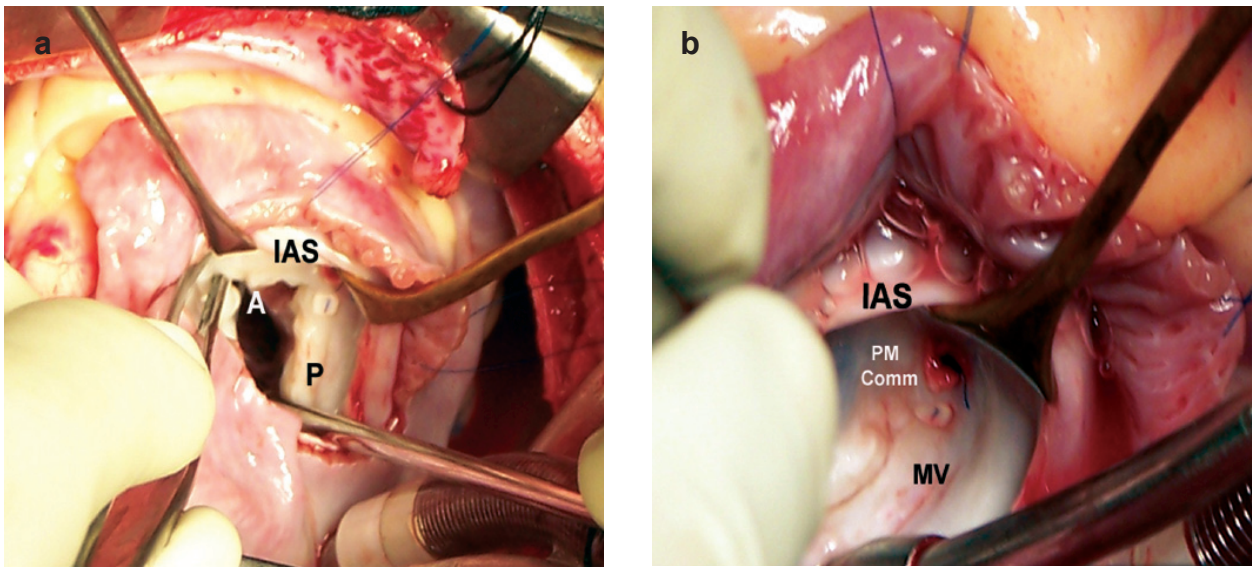

Figure 2. Intraoperative images: Thickened anterior (A) and posterior (P) mitral leaflets; repaired mitral valve (MV) with plicated posteromedial commissure (PM Comm); IAS = interatrial septum.

out.[1] Good functional competence of the mitral valve was restored (Figure 2b). The patient was weaned off cardiopulmonary bypass with satisfactory hemodynamics on inodilator support. Postoperative 2-dimensional echocardiography revealed trivial mitral regurgitation and a mitral valve area of 1.8 cm^2. She was discharged on decongestant medication, 3 weekly injections of 600,000 units of benzathine penicillin and sildenafil, which was continued for 3 months postoperatively. She is doing well at follow-up.

DISCUSSION

Rheumatic heart disease occurs as a late sequela of immune-mediated damage to the cardiac valves following throat infection with group A β-hemolytic *streptococci*.[2] An identical mechanism of injury to the developing fetal heart tissue by maternal antibodies produced in response to streptococcal throat infection has been hypothesized to explain the pathogenesis of hypoplastic left heart syndrome.[3] Congenital heart disease occurs secondary to developmental defects in the embryonal stage, with reported prevalence rates of up to 4.2 per thousand.[4] Rheumatic heart disease, a significant health problem in India due to the lack of a nationwide preventive program, is reported to be on the decline with a clinical prevalence of up to 4.54 per thousand and an echocardiographic prevalence of 0.68 per thousand.[5,6] Although it is not possible to determine whether the presence of RHD in congenital heart disease is a mere coincidence or if congenital heart disease actually predisposes to RHD, coexisting pathology has been described.[2,7]

Ventricular septal defect may coexist with congenital mitral stenosis, but the association of rheumatic mitral stenosis with multiple VSD has not been reported. Although congenital mitral stenosis may present with some commissural fusion and leaflet thickening, isolated lesions are usually severe and often produce symptoms and death during the first 4 to 5 years of life when left untreated. In the presence of important coexisting cardiac anomalies as in our case, symptoms are expected to occur even earlier.[8] Our assumption of rheumatic etiology based on echocardiography and the intraoperative appearance of a normally developed mitral valve apparatus is supported by a history of fever with migratory polyarthritis and survival of this child to the age of 7 years in the presence of mitral stenosis. This report highlights the need for careful evaluation of patients with congenital heart disease for a coexisting rheumatic condition, and attention to the possibility of a rheumatic process affecting them during subsequent follow-up.[7]

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