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Clinical case report based study

Juvenile severe mitral stenosis predisposing Eisenmenger syndrome in a case with ventricular septal defect, patent ductus arteriosus, coarctation of aorta & hypoplastic aortic arch: Report of first case of rare association

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ABSTRACT

We are reporting the first case of rare association between multiple congenital cardiac malformations with severe rheumatic mitral stenosis which is an acquired structural cardiac disease. A 16 years old female patient presented with progressive dyspnoea & cyanosis for the last one month with past history of recurrent pneumonia since infancy. Physical examination revealed presence of cyanosis, grade I clubbing, radio-radial & radio-femoral delay, loud & single second heart sound, apical long mid diastolic murmur and left parasternal ejection systolic murmur. Transthoracic echocardiography revealed severe rheumatic mitral stenosis, multiple ventricular septal defects (VSD) with bidirectional shunt, hypoplastic aortic arch, Coarctation of aorta and severe pulmonary hypertension. Transesophageal echocardiography revealed the same findings along with the presence of moderate mitral regurgitation and 9 mm peri-membranous VSD extending into muscular septum. Cardiac catheterization study confirmed the echo-cardiographic findings and demonstrated large patent ductus arteriosus (PDA). We have planned for high-risk percutaneous transmitral commissurotomy (PTMC) for this patient to decrease the back pressure on pulmonary vasculature. So that right to left shunt will be decreased and cyanosis will also improve. But parents refused to give consent for PTMC. She was on treatment with regular penicillin prophylaxis, diuretics, sildenafil and infective endocarditic prophylaxis. We should be aware of this kind of complex association between congenital and acquired structure heart disease. Eisenmenger syndrome could also be a presentation of juvenile severe rheumatic mitral stenosis when it is associated with congenital shunt lesion like VSD/PDA in our case.

1. Introduction

Rheumatic heart disease (RHD) still a major health problem in India and juvenile mitral stenosis is frequently seen in Indian subcontinent.[1] Congenital heart disease is related to events occurring in the embryonal stage, while rheumatic heart disease is an auto immune-mediated cardiac damage following streptococcal infection.[1] Though congenital heart diseases (CHD) like ventricular septal defect (VSD), patent ductus arteriosus (PDA), atrial septal defect (ASD) and bicuspid aortic valve can be associated with RHD,[2] but combination of different congenital heart defects like VSD, PDA, Coarctation of aorta (CoA) and type B hypoplastic aortic arch associated with juvenile mitral stenosis (MS) and presented with Eisenmenger syndrome is nowhere mentioned in the present literature.

2. Case report

A 16 year old female patient presented with progressive dyspnoea of New York Heart Association (NYHA) grade II–III & cyanosis for the last one-month. She had past history of recurrent pneumonia since infancy though she didn't give any history suggestive of rheumatic fever in past. She was stunted and wasted than her other siblings, though there was no family history of similar problem. General physical examination revealed presence of cyanosis, grade I clubbing, radio-radial & radio-femoral delay and significant blood pressure differences between right upper limb and left upper limb or both lower limb. Cardiovascular examination

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revealed loud first heart sound & single second heart sound, apical long mid diastolic murmur & grade II pansystolic murmur and left parasternal grade II ejection systolic murmur. Electrocardiogram showed extreme right axis deviation, biatrial enlargement, right ventricular hypertrophy (RVH) and strain pattern. Chest X-ray showed cardiomegaly with prominent pulmonary artery with peripheral pruning of pulmonary arteries. Transthoracic echocardiography (TTE) showed severe MS (mitral valve orifice area-0.9 mm^2 & mitral valve gradient-10/8 mm of Hg), submitral fusion (Fig. 1), grade I MR, multiple VSD with bidirectional shunt (muscular VSD – 7 mm; perimembranous VSD – 6 mm), hypoplastic aortic arch of type B, CoA, moderate tricuspid regurgitation and pulmonary hypertension (pulmonary artery systolic pressure – 85 mm of Hg). Transesophageal echocardiography (TEE) revealed the same findings along with the presence of moderate mitral regurgitation and 9 mm perimembranous VSD extending into muscular septum and valve suitable for percutaneous transmitral commissurotomy (PTMC) with Wilkins score of 5/16. Cardiac catheterization was done by using 6F Cournard (Medtronic) and 5F pigtail catheter (Cordis) and which showed RVH with dilated main pulmonary artery (MPA) and branch pulmonary arteries, type B hypoplastic aortic arch with absent left subclavian artery, CoA, large PDA and multiple VSD (Figs. 2 and 3). Findings of cardiac catheterization study before and 5 min after 100% O_2 administration confirmed the presence of Eisenmenger syndrome with little improvement in pulmonary-systolic shunt ratio (Qp/Qs), pulmonary vascular resistance index (PVRI) and pulmonary artery pressure (PAP). Due to non-improvement of PAP, Qp/Qs & PVRI after O2 therapy, surgical correction of VSD and PDA was not possible in our case. But as Eisenmenger syndrome was predisposed by juvenile MS in our case and mitral valve was also suitable for PTMC so only high risk PTMC was planned to decrease PASP and thus right to left shunt. But parents of the patient had refused to give consent for PTMC. So, she was put on sildenafil, diuretics, beta-blocker and three weekly intramuscular injection benzathine penicillin prophylaxes.

3. Discussion

The incidence of CHD is about 8 per 1000 live births. The prevalence of RHD in school-aged children is estimated to be in the range of 2–11 per 1000. Hence, the occurrence of both these conditions in the same patient can be considered to be uncommon.[1] In a previous study from Nepal, the prevalence of RHD and CHD among schoolchildren is 1.2/1000 and 1.3/1000, respectively, with mitral regurgitation and ASD being the commonest lesions.[3] In a study from India showed only few cases had both the diseases with ASD, VSD, PDA and bicuspid aortic valve being the most common congenital heart lesion.[2] They found that the prevalence of rheumatic fever/RHD was significantly higher in children with CHD (8.8%) as compared to those without CHD (0.3%).[2] Another study from India also showed the similar prevalence with 1.8% children with congenital or acquired heart disease who happened to have both types of lesions.[1] It is impossible to determine whether the presence of CHD with RHD is a mere coincidence or whether the presence of CHD actually predisposes to RHD.[4] There are few case reported in the literature where RHD with severe MS was associated with VSD.[5,6] Another case reported in 1956, which showed association between RHD and Eisenmenger syndrome.[7] But, our case was very peculiar because she had presented with the clinical features Eisenmenger syndrome as the primary

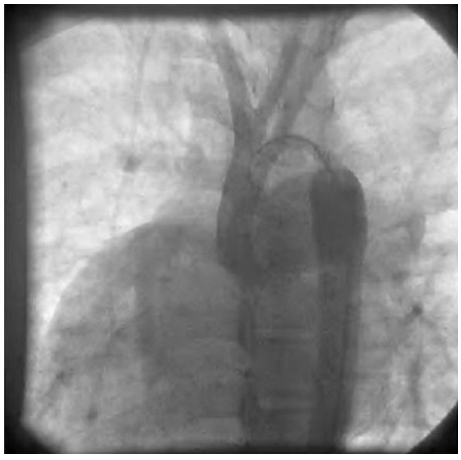

Fig. 2. Aortogram done by pigtail catheter showed Type B hypoplastic aortic arch with coarctation of aorta and absent left subclavian artery and post stenotic dilatation of descending thoracic aorta.

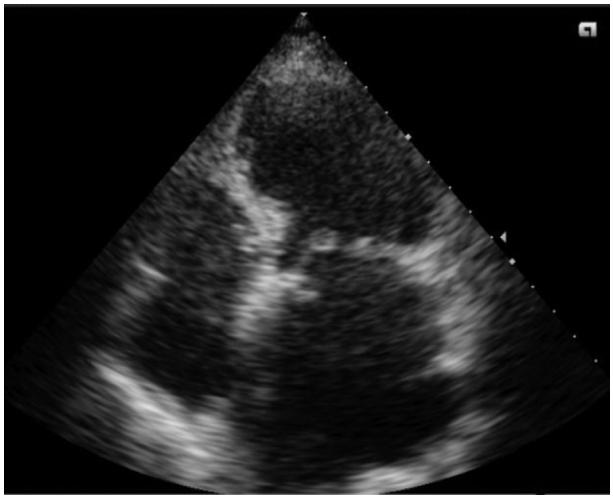

Fig. 1. Apical 4C view showed presence of rheumatic severe MS with thickened cusp.

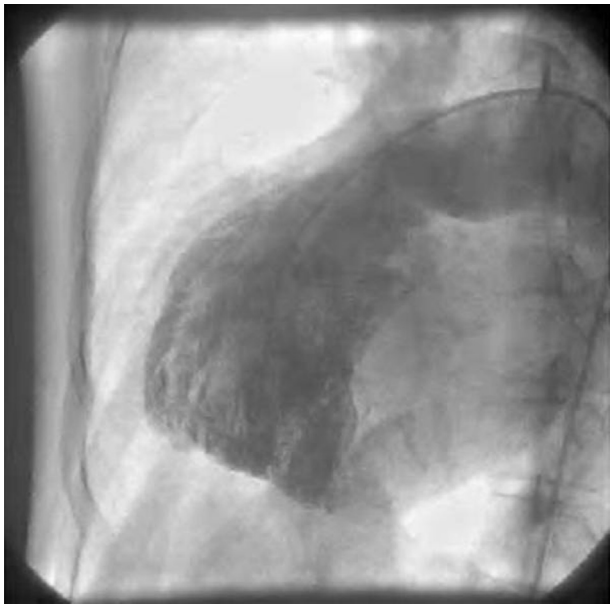

Fig. 3. RV angiogram done by Cournand catheter placed via the course of descending thoracic aorta ➔PDA➔pulmonary artery➔RV, showed hypertrophic RV, dilated PA and two VSD with staining of LV.

presentation for both RHD and VSD and in our case juvenile MS with severe MS was also predisposing the Eisenmenger syndrome due to the presence of multiple VSD & PDA. Type B hypoplastic aortic arch with CoA was also associated with this combination of CHD and RHD. Among the previous cases, both the cases had undergone surgical correction of VSD and MVR.[5,6] But, in our case surgical correction was not possible due to irreversibility of PAP & PVRI after O2 administration though PTMC could be done as mitral valve was suitable for it.[8] But due to refusal by parents we couldn't do the procedure in our case and she was managed medically only.

4. Conclusion

We should be aware of this kind of complex association between congenital and acquired structure heart disease. Eisenmenger syndrome could also be a presentation of juvenile severe rheumatic MS when it is associated with congenital shunt lesion like VSD/PDA in our case. This report highlights the need for careful evaluation of patients with CHD for a coexisting rheumatic heart diseases, and attention to the possibility of affecting with the rheumatic process during subsequent follow-up.

Contributors

SP, BK & SKS were involved in the management of this patient. SP & BK reviewed the literature & drafted the manuscript. SKS corrected the manuscript. All authors approved the final version of the manuscript.

Conflicts of interest

All authors have none to declare.

References

1. Bokhandi SS, Tullu MS, Shaharao VB, Bavdekar SB, Kamat JR. Congenital heart disease with rheumatic fever and rheumatic heart disease: a coincidence or an association? *J Postgrad Med*. 2002;48:238.
2. Gupta I, Gupta ML, Parihar A, Gupta CD. Epidemiology of rheumatic and congenital heart diseases in school children. *J Indian Med Assoc*. 1992;90:57–59.
3. Bahadur KC, Sharma D, Shrestha MP, et al. Prevalence of rheumatic and congenital heart disease in schoolchildren of Kathmandu valley in Nepal. *Indian Heart J*. 2003;55:615–618.
4. Befeler B, Hildner FJ, Cohen LS, Javier RP, Samet P. Combined rheumatic and congenital heart disease in adult patients. *J Fla Med Assoc*. 1971;58:24–27.
5. Sai Krishna C, Ramesh Babu V, Ram Mohan K, Baruah DK, Naresh Kumar PV. Juvenile rheumatic mitral stenosis with multiple ventricular septal defects. *Asian Cardiovasc Thorac Ann*. 2007;15:432–434.
6. Chiu CZ, Shyu KG. Coexistent supracristal ventricular septal defect and severe rheumatic mitral stenosis in a middle-aged woman: a case report. *FJJM*. 2009;7:39–45.
7. Mf Lion, Soberon Acevedo J. Eisenmenger complex associated with rheumatic heart disease. *Rev Paul Med*. 1956;48:237–249.
8. Rahimtoola SH, Durairaj A, Mehra A, et al. Current evaluation and management of patients with mitral stenosis. *Circulation*. 2002;106:1183–1188.