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Case Report Open Access

Supravalvular mitral ring in a patient with Turner syndrome: A case report

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ABSTRACT

Turner syndrome is a relatively common genetic disorder of female development characterized by partial or complete absence of an X chromosome. Some of the patients may present with cardiovascular abnormalities. However, there is no case with Turner syndrome with a supravalvular mitral ring (SMR) in the literature. Herein, we present a case of SMR accompanied by Turner syndrome who presented with progressive shortness of breath and exertional dyspnea, that was successfully treated.

Keywords: Mitral stenosis, supramitral ring, Turner syndrome.

Supravalvular mitral ring (SMR) is a rare anomaly of the mitral valve. It is a circumferential ridge or membrane that arises from the left atrial wall. The thickness and extent of the SMR may vary from a thin membrane to a thick discrete fibrous ridge.[1] The mitral valve is often abnormal and may be accompanied by valvular stenosis or fusion of leaflets, a small valve orifice, and abnormal papillary muscle morphology.[2] Adhesion to the mitral valve can disrupt its function, which is the major mechanism of mitral flow obstruction. It can progress over time and reduce cardiac output.[1] More than 90% of all SMR cases are associated with other congenital abnormalities,[1] and the incidence of concomitant SMR in Turner syndrome is unknown. Herein, we report a case of SMR accompanied by Turner syndrome who presented with progressive shortness of breath and exertional dyspnea that was successfully treated.

CASE REPORT

A 15-year-old female who was diagnosed with Turner syndrome presented to our clinic with exertional dyspnea and rapid fatigue (New York Heart Association [NYHA] Class III). The electrocardiogram showed sinus rhythm. The echocardiography revealed severe mitral stenosis due to supramitral ring. The peak and mean mitral valve gradients were 49 and 26 mmHg, respectively (Figure 1). The mitral valve diameter was 12 mm (Figure 2). The left atrium was dilated, and the interatrial septum was deviated to the right. The echocardiogram also showed a bicuspid aortic valve and a patent foramen ovale. The findings were confirmed by transesophageal echocardiography. There were no other pathologies which required an additional intervention. During surgery, the mitral valve was explored through a transseptal approach. A mitral ring was observed originating from the middle of the anterior mitral leaflet and extending toward the posterior leaflet and, then, to the supravalvular region (Figure 3). The tissue was excised. Any interchordal space obliterations were corrected (Figures 4 and 5). Follow-up surgical inspection revealed a good valve diameter. The postoperative transesophageal echocardiography showed a mean mitral valve gradient of 6 mmHg (Figure 6). The patient was discharged without any complications. A written informed consent was obtained from the parents and/or legal guardians of the patient.

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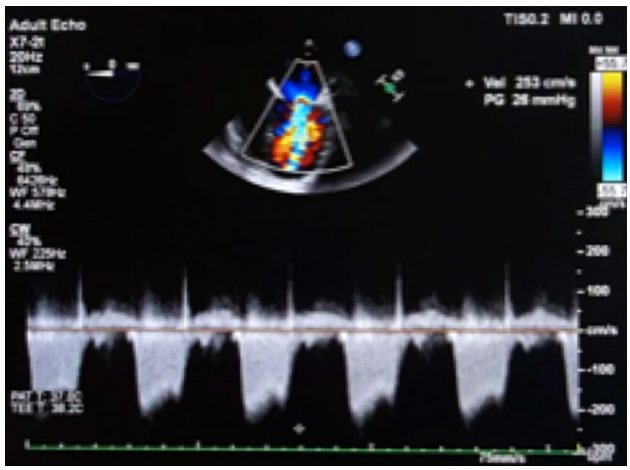

Figure 1. Preoperative mitral stenosis gradient.

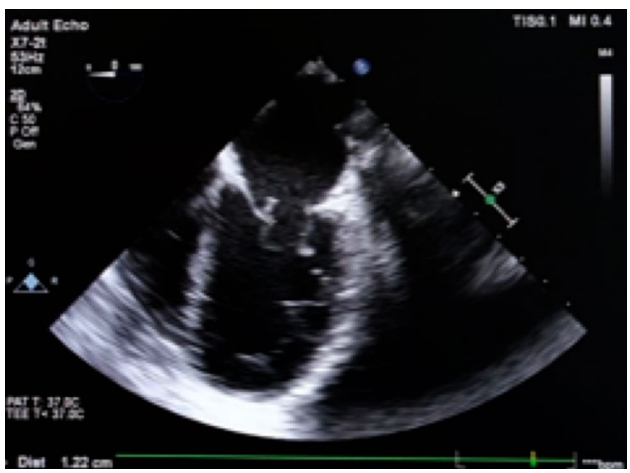

Figure 2. A preoperative view of mitral valve opening and supramitral ring.

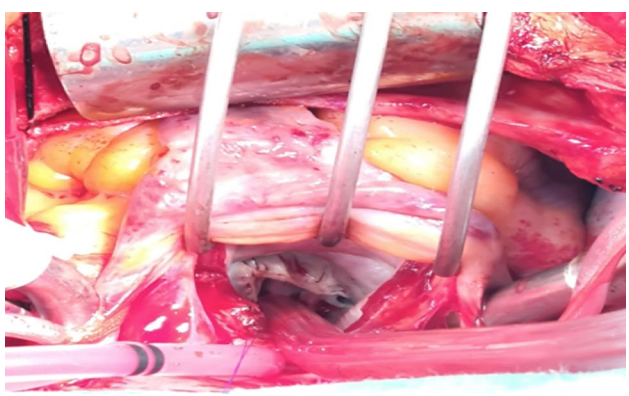

Figure 3. An intraoperative view of supramitral ring.

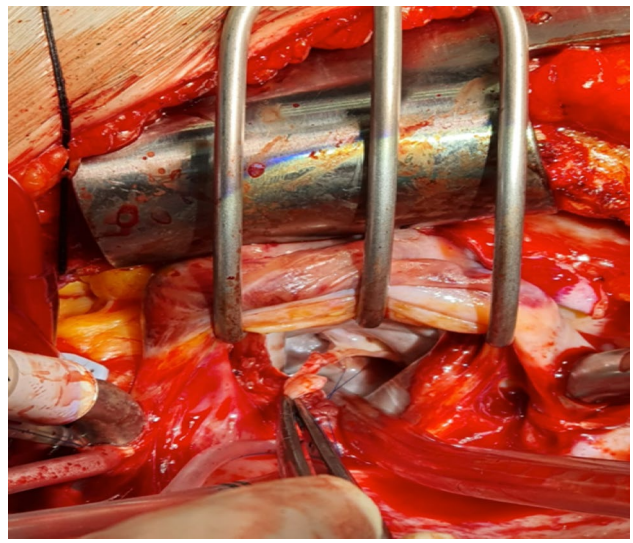

Figure 4. An intraoperative view of supramitral ring resection.

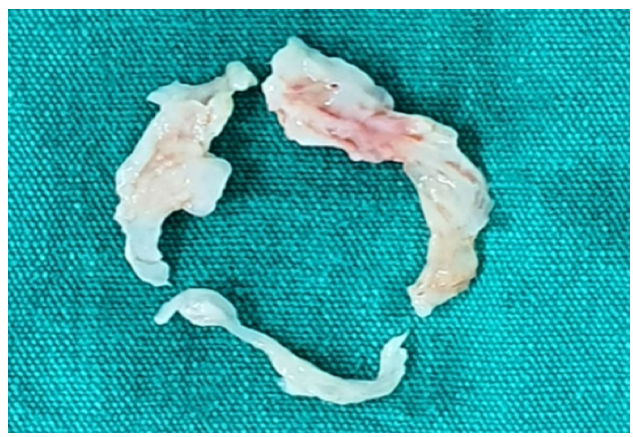

Figure 5. View of the ring after resection.

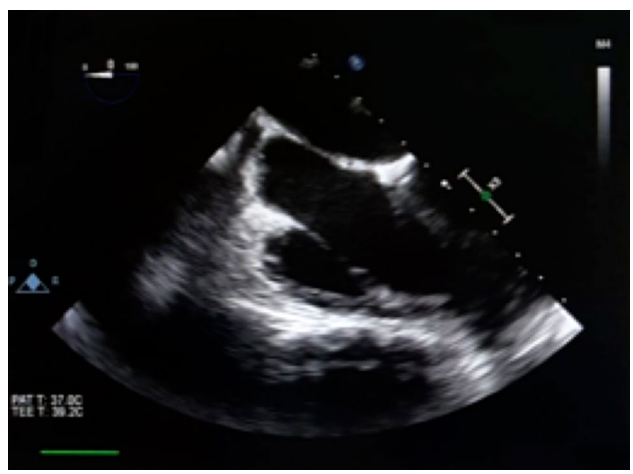

Figure 6. Postoperative follow-up echocardiography showing mitral valve patency.

DISCUSSION

Several studies have shown an increased incidence of mitral valve disease in patients with Turner syndrome; however, the study of Bondy et al.[3] found that the incidence of mitral valve disease was similar in Turner syndrome patients compared to the overall population.

Supravalvular mitral ring has not been described in patients with Turner syndrome. Supramitral ring rarely occurs as an isolated lesion and is often accompanied by other congenital heart diseases, including left-sided obstructive heart lesions, such as the parachute mitral valve, valvular and subvalvular aortic stenosis, bicuspid aortic valve, and coarctation of the aorta. It frequently presents as a part of Shone's complex. Supravalvular mitral ring has been rarely reported as concomitant with the Tetralogy of Fallot,[4] atrial septal defect, cor triatriatum, left-sided superior vena cava, unroofed coronary sinus, partial anomalous pulmonary venous return, pulmonary venous obstruction, double orifice mitral valve, transposition of the great arteries, atrioventricular discordance, or double outlet right ventricle.[5] Although cor triatriatum is a rare anomaly, it can be encountered in all age groups and should be considered in the differential diagnosis with supramitral ring, as both anomalies are similar in terms of clinical signs and symptoms.[6]

First described by Fisher[7] in 1902, SMR has been the subject of several studies to better understand the associated cardiac abnormalities. Collison et al.[8] proposed that cardiac anomalies associated with SMR could be classified into two main categories: SMR associated with ventricular septal defect, and SMR associated with left-sided obstructive pathologies, particularly sub-aortic membrane, bicuspid aortic valve, and coarctation of the aorta. In the first scenario, it is important to rule out SMR in the context of patients with ventricular septal defect and turbulence across the mitral valve. In the second scenario, it has been argued that supramitral ring should be excluded in patients with multi-level left heart obstructions to prevent residual defects.[8] In the light of these findings, the lesions described for the second scenario and the cardiovascular pathologies associated with Turner syndrome show more overlap. The present case had also a bicuspid aortic valve, but there was no obstructive lesion requiring an intervention.

A study by Toscano et al.[9] described two different variants of mitral ring, namely supramitral and intramitral. It is of surgical significance to differentiate these two subtypes of mitral pathologies. Supramitral ring is usually associated with a normal mitral valve apparatus and can occur concomitantly with congenital heart defects, as previously mentioned. Intramitral ring is usually a part of the Shone's complex and is associated with a normal subvalvular apparatus. The intramitral ring can, therefore, be considered a subtype of congenital mitral stenosis, characterized by varying degrees of obstruction, both at the level of the ring and the supravalvular apparatus. The supravalvular type is a form of left atrial obstruction, whereas the intravalvular type can be a part of an intrinsic mitral pathology.

Resection of the supramitral ring is simpler and more definitive, whereas intramitral ring resection is more challenging. Although ring resection is necessary, it is rarely sufficient, as the anatomy of the subvalvular apparatus significantly contributes to inflow obstruction. Satisfactory hemodynamic results can be achieved through papillary muscle splitting and correction of the interchordal space obliterations to increase the effective mitral orifice. Commissurotomy may postpone and avoid valve replacement. Ring resection provides excellent long-term results in all cases of supramitral ring and in half of the cases of intramitral obstruction. Residual mitral stenosis is present in the remaining half of patients with intramitral obstruction. This condition is always accompanied by an abnormal supravalvular apparatus or valve hypoplasia, and follow-ups reveal moderate-to-advanced valve deficiencies, despite reoperation. Predictors of poor surgical outcomes include mitral annulus hypoplasia and abnormal supravalvular apparatus. Moreover, being younger than one year of age has been shown to be associated with poor surgical outcomes.[9] In their study, Yildirim et al.[10] argued that, even in patients with mitral annulus hypoplasia, the stenosis gradient could be reduced to reasonable values only with effective ring resection. Our case had no major pathology related to the subvalvular apparatus, and the existing interchordal obliterations were corrected.

The morphology and optimal treatment of the SMR are still controversial. Once detected, detailed echocardiography should be performed considering possible concomitant pathologies. In the presence of suspicious findings, the diagnosis should be confirmed with transesophageal echocardiography. A transseptal surgical approach can achieve optimal resection of the ring without causing mitral valve dysfunction, as

was the case with the patient in the present report. A transseptal surgical approach is also favorable, as it facilitates the detection of concomitant pathologies.[8]

In conclusion, although rare, supramitral ring should be considered in patients with Turner syndrome. Concomitant pathologies must be carefully considered before surgery. A transseptal surgical approach is recommended. Simple resection is sufficient for isolated lesions. Any concomitant mitral inflow obstruction must be corrected simultaneously. Early results are excellent for isolated defects.

Declaration of conflicting interests

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REFERENCES

1. Baharestani B, Sadat Afjehi R, Givtaj N, Sharifi M. Supravalvar mitral ring: A case report. J Tehran Heart Cent 2012;7:82-4.
2. Serra W, Testa P, Ardissino D. Mitral supravalvular ring: A case report. Cardiovasc Ultrasound 2005;3:19.
3. Bondy CA. Congenital cardiovascular disease in Turner Syndrome. Cong Heart Dis 2008;3:2-15.
4. Barik R, Nemani L. Tetralogy of Fallot associated with supramitral ring: "Paying for a near miss". J Cardiol Cases 2015;12:159-61.
5. Nair AK, Chowdhuri KR, Radhakrishnan S, Iyer KS, Saxena M. Rare combination of pathologies causing mitral stenosis and mitral regurgitation: A case report. World J Pediatr Congenit Heart Surg 2020;11:NP113-NP116.
6. Ozyuksel A, Yildirim O, Avsar M, Hayirlioglu M, Demiroluk S, Kucukosmanoglu O, et al. Surgical correction of cor triatriatum sinister in the paediatric population: Mid-term results in 15 cases. Eur J Cardiothorac Surg 2015;47:e25-8.
7. Fisher T. Two cases of congenital disease of the left side of the heart. Br Med J 1902;1:639-41.
8. Collison SP, Kaushal SK, Dagar KS, Iyer PU, Girotra S, Radhakrishnan S, et al. Supramitral ring: Good prognosis in a subset of patients with congenital mitral stenosis. Ann Thorac Surg 2006;81:997-1001.
9. Toscano A, Pasquini L, Iacobelli R, Di Donato RM, Raimondi F, Carotti A, et al. Congenital supravalvar mitral ring: An underestimated anomaly. J Thorac Cardiovasc Surg 2009;137:538-42.
10. Yildirim O, Ozyuksel A, Avsar M, Yerebakan C, Zeybek C, Demiroluk S, et al. Mid-term results of congenital supravalvar mitral ring resection. J Card Surg 2015;30:591-4.