Supratricuspid obstructive membrane in congenitally corrected transposition of the great arteries

Sanjeev Kumar, Milind Phadke, Prafulla Kerkar
Department of Cardiology, Seth G S Medical College and K E M Hospital, Mumbai, Maharashtra, India

ABSTRACT

Obstructive lesions in the inflow of the systemic ventricle in congenitally corrected transposition of the great arteries are rare. It is important to identify such lesion which could alter the surgical outcome if not recognized. We report the echocardiographic findings in a patient with supratricuspid obstructive membrane with corrected transposition of the great arteries.

Keywords: Corrected transposition of great arteries, echocardiography, supratricuspid membrane

An 18-month-old male child was referred to our institute with complaints of fast breathing and repeated episodes of lower respiratory tract infections requiring hospitalizations in the past. On examination, he was underweight with no evidence of cyanosis or clubbing. There was a pansystolic murmur of grade III/VI along the lower left sternal border. The arterial oxygen saturation was 99%. His chest X-ray showed levocardia, cardiothoracic ratio of 0.6 with a left ventricular type apex and plethoric pulmonary vasculature. The echocardiogram was suggestive of levocardia, situs solitus, and congenitally corrected transposition of the great arteries (CCTGA) with SLL loop. There was large subpulmonic ventricular septal defect with inlet extension and left to right shunting. There is no left ventricular outflow tract obstruction [Figure 1a]. In addition, an echogenic membrane was noted just above the left-sided atrioventricular valve (morphological tricuspid valve) and below the left atrial appendage in the modified apical four-chamber view. The valve and subvalvular apparatus were structurally normal with a valvular annulus of 15 mm (Z value = –1.12). The membrane was causing severe obstruction of right ventricular inflow with pressure gradients of 26/18 mm of Hg [Figure 1b and c]. Surgical correction with resection of membrane and closure of ventricular septal defect was advised but declined by the family.

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DISCUSSION

CCTGA accounts for less than 1% of the cases of congenital heart diseases. CCTGA is known to be associated with ventricular septal defect, atrial septal defect, and obstruction in right ventricular outflow tract, tricuspid valve anomalies, and coarctation of the aorta. Abnormalities of the morphological tricuspid valve (left atrioventricular valve in SLL loop) are frequently found during autopsy in CCTGA,[1] These abnormalities commonly resemble the Ebstein anomaly of a right-sided tricuspid valve in hearts without ventricular inversion.[2] Occurrence of obstructive lesions of right ventricular inflow is rare. A review of the literature revealed only a few individual case reports and a single autopsy series. Allwork *et al.*,[2] in their autopsy study of 32 cases of CCTGA, noted abnormalities of the morphological tricuspid valve in 91% of the cases; most of them were Ebstein malformations and only five had supravalvular stenosing ring. Marino *et al.*,[1] in their echocardiographic study of 42 cases of CCTGA, found only two cases with obstruction in right ventricular inflow. Both the cases also had subpulmonary obstruction. Supratricuspid membrane often originates from the left atrial wall below the left auricle, rarely from the tricuspid annulus. The pathophsyiological consequences of such a membrane are similar to the supramitral ring in hearts with concordant atrioventricular and ventriculoarterial connections. Both supratricuspid and supramitral are believed to develop after incomplete division of endocardial cushion tissue.[3] Toscano *et al.*, classified the supramitral ring into two categories based on the involvement of the valve: Supramitral type and intramitral

Address for correspondence: Dr. Sanjeev Kumar, Department of Cardiology, Seth G S Medical College and K E M Hospital, 4th Floor, CVTC Building, Parel, Mumbai - 12, Maharashtra, India. E-mail: dr_rahulreddy@yahoo.co.in

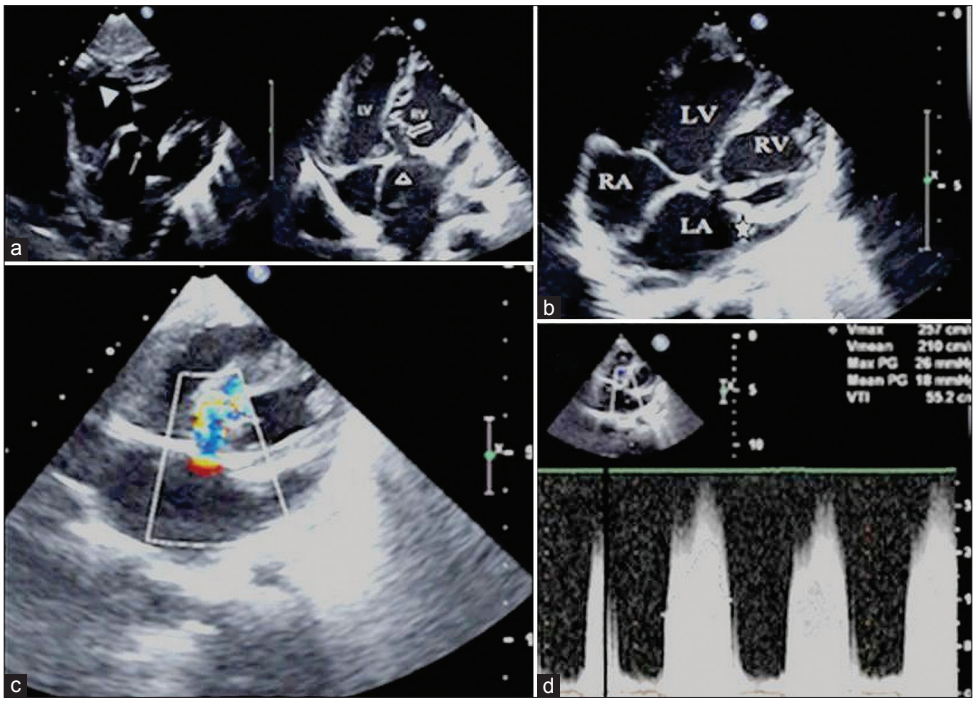

Figure 1: (a) Echocardiographic apical four-chamber view showing mitral (arrow) and pulmonary valve (arrow head) continuity with apical displacement of the septal leaflet of the tricuspid valve which is trying to restrict the ventricular septal defect (open arrow); the supratricuspid membrane is separately seen (open arrow head); the membrane (marked with star in panel b) is distinctly visualized in the modified apical four-chamber view; panel c shows turbulent flow across the membrane with gradients of 26/18 mm of Hg (panel d)

type. In the intramitral type, the membrane is attached to the mitral valve and is associated with the involvement of the subvalvular apparatus. The distinction between the two types is important, as the surgical approach and outcome differ.[4] Collison *et al.*, in their surgical case series did not report recurrence of the supramitral ring after a mean follow-up of 30 months.[5] However, Toscano *et al.*, reported the need for reoperation in four of eight patients with intramitral-type membrane over a mean follow-up of 21.5 months. In the present case, the membrane falls in the true supravalvular category. In addition to other associated lesions, it is important to recognize this particular abnormality, as uncorrected obstruction in the inflow can lead to pulmonary venous hypertension[6] and may necessitate an alteration in the surgical plan. Echocardiography is an excellent noninvasive tool for diagnosis of this rare entity and obviates the need for catheterization.

REFERENCES

1. Marino B, Sanders SP, Parness IA, Colan SD. Obstruction of right ventricular inflow and outflow in corrected transposition of the great arteries (SLL): Two-dimensional echocardiographic diagnosis. J Am Coll Cardiol 1986;8:407-11.
2. Allwork SP, Bentall HH, Becker AE, Cameron H, Gerlis LM, Wilkinson JL, *et al.* Congenitally corrected transposition of the great arteries: Morphologic study of 32 cases. Am J Cardiol 1976;38:910-23.
3. Choi ES, Kim WH, Park SJ, Min BJ. Right ventricular inflow obstruction caused by supratricuspid ring after the conventional biventricular repair of congenitally corrected transposition of great arteries. Korean J Thorac Cardiovasc Surg 2011;44:58-60.
4. 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.
5. 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.
6. Gupta V, Kothari SS, Sharma R, Bhan A. Systemic inflow obstruction in congenitally corrected transposition of great arteries. Indian Heart J 1997;49:81-2.

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