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Right Ventricular Inflow Obstruction Caused by Supratricuspid Ring after the Conventional Biventricular Repair of Congenitally Corrected Transposition of Great Arteries

— A case report —

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A seventeen-month-old male baby, who had received conventional biventricular repair for congenitally corrected transposition of the great arteries, underwent excision of supratricuspid ring. Although tricuspid valve annulus was marginally small on direct inspection in the operating theater, circumferential excision of supratricuspid ring alone completely relieved the right ventricular inflow obstruction.

Key words: 1. Congenital heart disease (CHD)
2. Stenosis
3. Transposition of great vessels

CASE REPORT

A one-month-old male baby, who had been born at 38^{+5} weeks of gestation with a birth weight of 3,700 gm, underwent conventional biventricular repair for congenitally corrected transposition of great arteries, ventricular septal defect (VSD), atrial septal defect (ASD), and patent ductus arteriosus (PDA). The surgical intervention consisted of trans-atrial-transmitral closure of VSD, direct closure of ASD and ligation of PDA. On echocardiography at postoperative 10 days, there was no residual left-to-right shunt, and the tricuspid valve showed mild regurgitation without inflow stenosis. He was discharged home, and began to be followed up at the outpatient department. At 17^{th} month old, he was readmitted for the evaluation of right ventricular inflow obstruction. Echocardiography revealed a significant pressure gradient between the left atrium (LA) and the right ventricle (RV) (15 mmHg), which was mainly caused by supratricuspid ring structure (Fig. 1). The patient underwent complete excision of the supratricuspid ring, which was almost obstructing the right ventricular inflow (Fig. 2, 3). Although the actual orifice size of the tricuspid valve measured at the operating theater was smallish (11 mm, Z valve=−3.5), surgical intervention for the tricuspid valve was deemed unnecessary because the morphology and function of the tricuspid valve appeared to be normal. Cardiopulmonary bypass time and aortic cross-clamping time were 89 minutes and 35 minutes, respectively. The LA-RV pressure gradient decreased to 3 mmHg immediate postoperatively. The patient has been closely followed up postoperatively because trans-tricupsid pressure gradient increased to 10 mmHg and the size of the tricuspid valve was still significantly small (11.9 mm, Z value:

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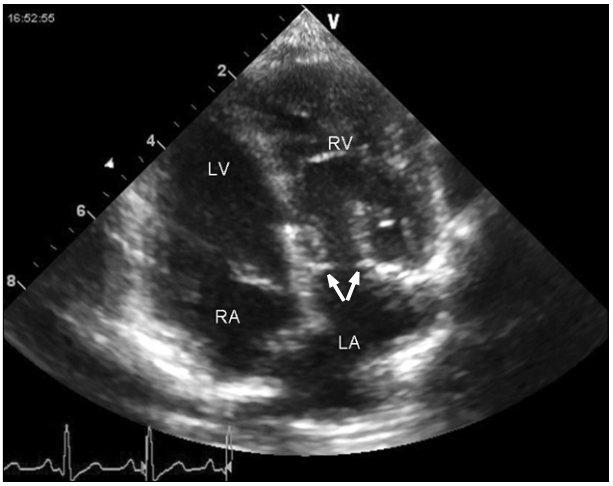

Fig. 1. Preoperative echocardiogram showing supratricuspid ring (white arrows). LA=The left atrium; RA=The right atrium; LV=The morphologic left ventricle; RV=The morphologic right ventricle.

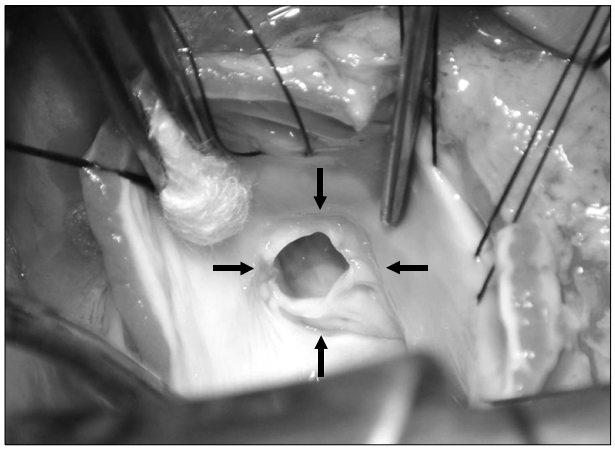

Fig. 2. Supratricuspid ring (Black arrows) observed through the left atriotomy.

−7) on follow-up echocardiography at postoperative 5 months.

DISCUSSION

Supratricuspid ring (STR) is a rare pathological condition, usually associated with congenitally corrected transposition of the great arteries (cc-TGA) [1,2]. Because this fibrous structure is originating from the left atrial wall, not from the atrioventricular valves, the pathogenesis of STR may well be

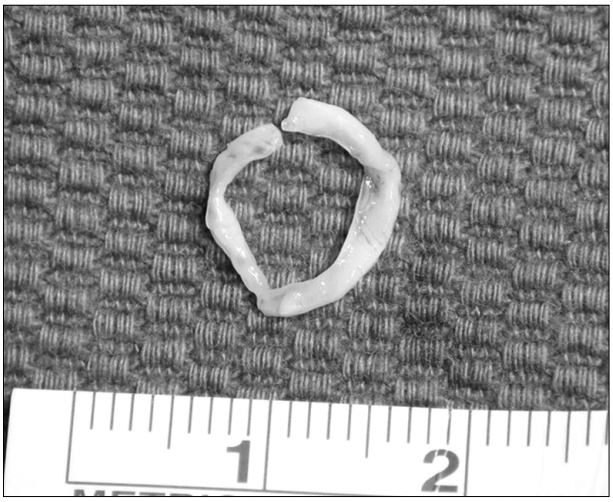

Fig. 3. Excised specimen of supratricuspid ring.

identical to supramital ring [3-5]. Both rings are believed to develop after the incomplete division of the endocardial cushion tissue, and, therefore, pathologic findings of the rings (un-laminated fibrous degeneration) are also likely to be the same. Cc-TGA is more frequently associated with VSD, ASD, or right ventricular outflow obstructrion (RVOTO), and the association of right ventricular inflow obstruction with cc-TGA is very rare. Once developed, STR in cc-TGA does not exist alone, and is frequently associated with aortic coarctation, subaortic stenosis, VSD, Ebstein's anomaly and RVOTO [1,2]. Accurate diagnosis is obtained by echocardiography, which shows bright echogenic membranous tissue right above the tricuspid valve and below the left atrial auricle. In cor triatriatum, which is one of the most important differential diagnoses of SMR, the obstructing membrane exist proximal to (or above) the left atrial auricle [1]. Because information on the treatment of STR is sparse and pathophysiology is similar to supramitral ring, surgical techniuqe for the latter can be referred to [3-5], except that surgical approach for the STR would well be through the left atriotomy while that for the supramitral ring is generally through the atrial septal incision. Information on the tricuspid valve pathology in association with STR is also limited. With respect to supramitral ring, Toscano et al. [3] classified supramitral obstructing structure into two discrete categories: supramital type and intramitral type. While supramitral type is not accompanied by mitral valve pathology, intramitral type was

defined as membranous tissue being attached to the mitral valve in association with the pathologic conditions of subvalvular apparatus. Thus, they claimed, surgical technique for intramitral type should include "peeling the membrane off the mitral valve" and aggressive intervention for the mitral valve pathology. To the contrary, Konstantinov et al. asserted that surgical intervention for the mitral valve in supramitral ring had no impact on the surgical outcome, except for the papillary muscle splitting for parachute mitral valve [4]. In our case, we elected not to intervene surgically in the tricuspid valve, because valve leaflet and subvalvar apparatus were deemed normal. Although prognosis of STR is unclear, long-term outcome is thought to be determined mainly by associated cardiac anomalies. When surgical outcome of supramitral ring is referred to, most of the ring structure is surgically resectable, and, after extensive excision, early and long-term outcome is reported to be better than other anomalies causing left ventricular inflow obstruction. Accoding to the report by Collison et al. [5], 15 patients underwent surgical resection of supramitral ring, and no patients among 14 survivors developed recurrent of left ventricular inflow obstruction during the mean follow-up of 30 months. Toscano et al, however, reported that, while no patient with supramitral type membrane developed recurrent left ventricular inflow stenosis, four out of eight patients with intramitral type membrane received reoperations for recurrent left ventricular inflow obstruction at the mean postoperative duration of 21.5 months [5]. They attributed congenital hypoplasia of the mitral valve and abnormalities of subvalvar apparatus to poor clinical outcome. Based on the Toscano's classification [3], our case could be categorized as supratricuspid type, rather than intra-tricuspid type, but the patient developed moderate left ventricular inflow obstruction 5 months after the resection, presumably due to annular hypoplasia of the tricuspid valve. Therefore, careful inspection of the tricuspid valve and aggressive surgical intervention, if tricuspid valve annulus is hypoplastic, are mandatory upon the resection of supratricuspid ring associated with cc-TGA.

REFERENCES

1. Marino B, Sanders SP, Parness IA, Colan SD. *Obstruction of right ventricular inflow and outflow in corrected transposition of the great arteries {S,L,L}: two-dimensional echocardiographic diagnosis.* J Am Coll Cardiol 1986;8:407-11.
2. Chesler E, Beck W, Barnard CN, Schrire V. *Supravalvular stenosing ring of the left atrium associated with corrected transposition of the great vessels.* Am J Cardiol 1973;31: 84-8.
3. Toscano A, Pasquini L, Iacobelli R, et al. *Congenital supravalvar mitral ring: an underestimated anomaly.* J Thorac Cardiovasc Surg 2009;137:538-42.
4. Konstantinov I, Yun TJ, Calderone C, Coles JG. *Supramitral obstruction of left ventricular inflow tract by supramitral ring.* Oper Tech Thorac Cardiovasc Surg 2004;9:247-51.
5. Collison SP, Kaushal SK, Dagar KS, et al. *Supramitral ring: good prognosis in a subset of patients with congenital mitral stenosis.* Ann Thorac Surg 2006;81:997-1001.