Left Ventricular-Right Atrial Communication with an Aneurysm of the Atrioventricular Membranous Septum

A Case Report

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SUMMARY

A 40-year-old man with combination of left ventricular-right atrial communication and an aneurysm of the atrioventricular membranous septum is presented. A supravulvar type defect, 14×15 mm in size, was closed with a Teflon patch. An oval shaped aneurysm of the atrioventricular membranous septum, 20×17 mm in size was also noted, but was left untouched. To our knowledge, this is the first reported case with this combination in Japan.

Additional Indexing Words:

Left ventricular-right atrial communication Aneurysm of the
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A congenital heart anomaly with a shunt from the left ventricle to the right atrium is generally termed a left ventricular-right atrial communication (LV-RAC) and is divided into supravulvar and infravulvar types, depending on the site of communication. Thurnam¹⁾ originally described the disease in 1838, Buhl²⁾ was the first to describe a true atrioventricular communication, and Kirby et al,³⁾ were the first to make a successful radical repair in 1957. Although it is rather rare, several other cases have been reported.⁴⁾⁻¹³⁾

According to Lev and Shapir,¹⁴⁾ the first description of an aneurysm of the membranous septum was, attributed to Laennec in 1826 and first case was

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diagnosed clinically by Steinberg¹⁵⁾ in 1957. This condition is also considered to be uncommon. Recently, we have treated a patient with both LV-RAC and an aneurysm in the atrioventricular membranous septum. This communication is to report our experiences in detail.

CASE REPORT

A 40-year-old man was admitted to our hospital for examination of a heart murmur. His chief complaint was easy fatigability. There was no history of frequent respiratory infections, bacterial endocarditis or arrhythmia. There was a palpable vibration at the left sternal border in the 4th intercostal space. A rough and low-pitched grade 4/6 systolic murmur was heard over the entire precordium, and was loudest at the apex. The chest roentgenogram showed moderate cardiac enlargement, involving principally the right atrium and the right ventricle. Left ventricular hypertrophy, a high amplitude, peaked P wave and incomplete right bundle branch block were observed electrocardiographically. Cardiac catheterization showed an elevated mean right atrial pressure of 15 mmHg and pulmonary arterial pressure of 40/20 mmHg.

Fig. 1. Angiography. A: Pre-operative left ventriculography. B: Post-operative left ventriculography.

Fig. 2. Diagram of surgical procedure.

A left-to-right shunt of 60% was also demonstrated at the atrial level. Left ventriculography revealed immediate opacification of the right atrium (Fig. 1A).

The patient underwent open heart correction on July 18, 1978, under the suspicion of LV-RAC. Through mediansternotomy, enlargement of the right atrium and right ventricle was observed. A systolic vibration was palpable over the right atrial wall near the ascending aorta. Manual exploration of the right atrial cavity through the right atrial appendage revealed a stream of blood from the left ventricle into the right atrium, and the diagnosis of LV-RAC was confirmed. When the atrium was opened, bright red blood spurted out through the communication. The defect, a supravulvar type (14x15 mm in size) was closed by a Teflon patch. An oval shaped aneurysm (20x17 mm in size) was found by careful examination of membranous septum in the atrium. The aneurysm was posterocranial to the communication. It was left untouched (Fig. 2). The postoperative course was uneventful. Cardiac catheterization performed 2 months following surgery showed a significant reduction of the right atrial pressure to 3 mmHg and no residual shunt. Left ventriculography showed no residual shunt, but only the aneurysm in the membranous septum (Fig. 1B).

DISCUSSION

The observation of LV-RAC with an aneurysm of the membranous sep-

tum is rare. Sakakibara¹⁶⁾ reported only 13 cases with LV-RAC out of his 6,721 cases with congenital heart disease. In our literature survey of supra-valvular LV-RAC, there are 8 reported^{5)-8),11)-13),17)} cases in Japan, although details of 2 of these cases^{5),7)} were not documented. The current case is the ninth and is probably the first associated with an aneurysm of the membranous septum. Cardiac catheterization showed a definite rise of O₂ saturation in the right atrium in 5 cases, and in the right atrium and right ventricle in 1 case. Left ventriculography was performed in only 2 of 7 cases. Additional malformations occurred in about one third of the cases. An atrial septal defect is the most common associated lesion,⁹⁾ and an aneurysm of interventricular membranous septum, ventricular septal defect, atrial septal defect, coarctation of the aorta, high origin of the coronary artery, and aortic valve cusp deformity, are the common associated cardiac lesions.^{2),18)} However, the combination of an aneurysm of the atrioventricular membranous septum and supra-valvular LV-RAC has not been found.¹⁸⁾⁻²⁰⁾

There are several theories^{19),21)-23)} regarding the development of the aneurysm of membranous septum. According to Lev and Saphir,¹⁴⁾ an embryologic basis for the formation of a membranous septal aneurysm may be provided by either a defective formation of the endocardial cushion or an abnormality in the fusion of the caudal ends of the truncocoanal septum and the muscular ventricular septum. The latter explanation, first proposed by Mall,²¹⁾ was favored by Lev and Saphir. The most recent explanation proposed is that the aneurysm of membranous septum results from a previous, spontaneously closed ventricular septal defect and a high left ventricular pressure, which causes bulging to the right of an intrinsically weak membranous septum.^{18),20),24),25)} Because our case does not conform to Mall's anatomical observations or a ventricular septal defect, abnormalities in fusion of the endocardial cushion could be considered as a probable cause.

As to the surgical treatment for an aneurysm of the membranous septum, Vidne and associates¹⁸⁾ reported 29 cases in which complete correction was performed and proposed that, in the presence of an aneurysm, even a small ventricular septal defect should be closed by a patch and the aneurysm should perhaps be excised. Honda and colleagues²⁶⁾ reported 12 cases which were treated surgically by imbrication in the suture line of the ventricular septal defect, imbrication with a Teflon patch or simple patching with Teflon. A long-term follow-up of these patients would be needed to evaluate the surgical effect on aneurysms of the membranous septum.

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