Yamanashi Med. J. 5(1), 75~78, 1990

Case Report

A Rare Case of Congenital Downward Displacement of Right Aortic Annulus Resulting in Severe Aortic Regurgitation

Tetsunosuke MATSUKAWA, Shinpei YOSHII, Ryoichi HASHIMOTO, Tatsuro MAEDA[1)], Tetsuyo YOSHIZAKI[1)], Koji TAMURA[1)], and Akira UENO

Second Department of Surgery, Yamanashi Medical College, [1)]Second Department of Internal Medicine, Yamanashi Medical College, Tamaho, Makakoma, Yamanashi 409–38, Japan

Abstract: Isolated congenital aortic regurgitation in the three-cusped aortic valve is extremely rare. A 45-year-old man underwent successful aortic valve replacement for severe aortic regurgitation, resulting from the downward displacement of the right aortic annulus with a hypoplastic cusp, which originated from the interventricular septum. Its origin was considered to be congential because of its anatomical rarity.

Key words: downward displacement, aortic annulus, congenital, aortic regurgitation

Isolated congenital aortic regurgitation is considered very rare[1)]. It is less commonly caused by bicuspid or more rarely by quadricuspid aortic valve dysfunction without stenosis. In regard to the three-cusped aortic valve, only a few cases have been reported. Of these, one of the three aortic cusps was dysplastic and attached to the aortic wall[2–4)]. Another was distorted by accessory tissue[5)] resulting in pure aortic regurgitation. And recently, a very rare case of downward displacement of the aortic annulus of congenital origin has been reported[6)]. In this report, we describe an unusual case of severe aortic regurgitation resulting from the downward displacement of the right aortic annulus, which originated from the interventricular septum.

CASE REPORT

A 45-year-old man was admitted to the Yamanashi Medical College Hospital with a five year history of exertional palpitations, chest pain and pretibial edema. He had had no previous episode of rheumatic fever or infective endocarditis, but a cardiac murmur without any causal symptoms had been noted since early childhood.

Received, October 28, 1989
Accepted, November 22, 1989

On admission in April 1985, he was 173 cm in height and 69 kg in weight with no sign of marfan's syndrome or other connective tissue diseases. Physical examination showed a blood pressure of 152/40 mmHg and a regular heart rate of 58 per minute. Auscultation disclosed a grade II systolic ejection and a grade IV diastolic blowing murmur, with a "cooing" musical component at the third intercostal space of the left sternal border. Chest X-rays revealed mild cardiomegaly with left ventricular enlargement. An electrocardiogram showed sinus rhythm, a QRS axis of +30° and moderate left ventricular hypertrophy with ST-T depression in the II, III and aVF leads. Long-axis views of two-dimensional echocardiograms suggested that a small hyper-echoic right aortic cusp was displaced beneath the aortic annulus during the whole diastolic phase (Fig. 1-A to B). The left ventricular cavity was extremely dilated with an aortic annulus 28 mm in diameter. Its function was within

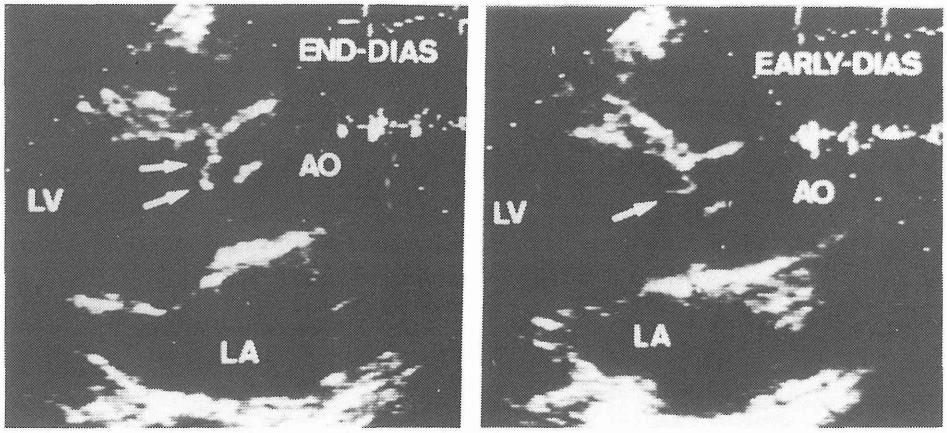

Fig. 1. Echocardiographic long-axis views of the heart; An enigmatical movement of the right coronary cusp (white arrow) of the aortic valve. A right coronary cusp seems to originate from the true annulus suggesting a cusp prolapse in this figure.

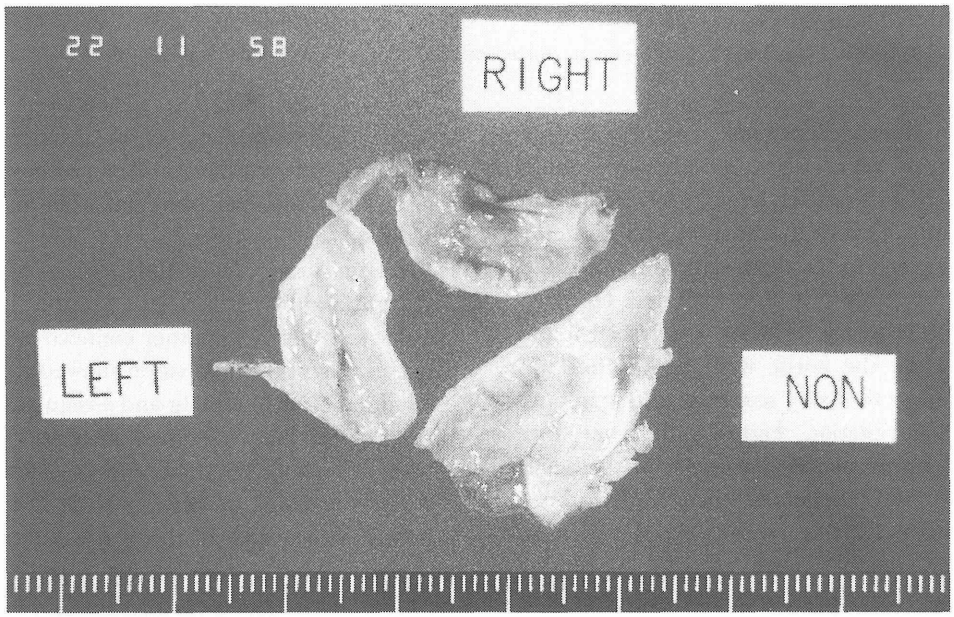

Fig. 2. Three excised coronary cusps; Left and non-coronary cusps were almost normal in size and nature. The right coronary cusp was thickened and smaller in size.

normal limit. There was no pressure gradient across the aortic valve. An aortic root angiogram demonstrated a grade IV aortic regurgitation, but no detailed aortic cusp abnormality appeared. there was no annulo-aortic ectasia. A coronary angiogram showed a wave-like deformity of the right coronary artery. These results suggested an initial diagnosis of severe aortic regurgitation primarily originating from a prolapse of the right coronary cusp. Later, laboratory data showed severe proteinuria resulting in hypoproteinemia (serum protein content was 3.8g/dl) which suggested "nephrotic syndrome" possibly of cardiac origin. The patient was functionally in NYHA class III.

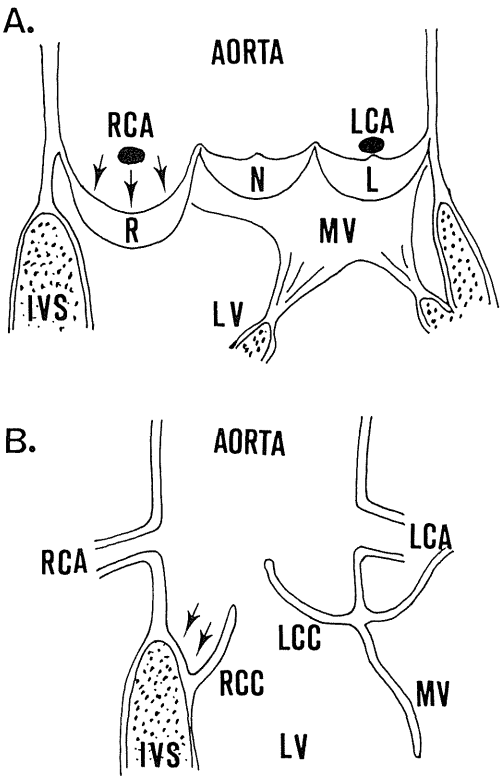

Fig. 3. Anatomical scheme of an aortic valve according to operative findings. A small right coronary cusp was grossly displaced into the left ventricle and originated from the interventricular septum (black arrow).
IVS, interventricular septum. LCC(L), left coronary cusp. LV, left ventricle. MV, anterior leaflet of mitral valve. RCC(R), right coronary cusp. LCA, left coronary artery. RCA, right coronary artery.

In October 1985, he underwent a conventional cardiopulmanary bypass. An oblique aortotomy was performed. The right coronary cusp of the valve was small and slightly thickened, and sunken excessively into the left ventricle. Further, it rose abnormally from the interventricular septum, distinctly below the normal position of the aortic annulus. The right coronary ostium was positioned normally, and the other two cusps (noncoronary and left) were almost normal in size and nature (Fig. 2). The right aortic cusp had a fixed movement both in systole and diastole in the left ventricle, resulting incomplete loss of incompetence aortic valve was replaced by a Bjork-Shiley prosthetic valve (size 27 A). The prosthesis was seated at the normal site of the aortic ring, above the attachment of the original aortic valve. A histological examination of the excised aortic cusps revealed no evidence of postinflammatory changes, but mild myxomatous degeneration was observed only in a noncoronary cusp.

Additionaly, acute nonoliguric renal failure developed, which was effectively treated by peritoneal dialysis in the immediate postoperative period. The serum protein content had increased to 6.3 g/dl 6 months later. The patient is doing well four years postoperatively.

Comment

Several causes of aortic regurgitation have been discussed, but congenital aortic regurgitation resulting from an isolated lesion is extremely rare[1–5]. In our patient, the attachment of the smaller right coronary cusp was grossly displaced downwards onto the interventricular septum, resulting in severe aortic regurgitation. The primary cause of regurgitation was considered to be congenital for the following reasons: the unusual appearance of the cusp, no postinflammatory changes of the excised right aortic cusp and the detection of a cardiac murmur since early childhood. However, the possibility of a non-congenital cause could not be ruled out completely, because exact pathoanatomical examination could not be performed in this case.

Levine and Harvey[7], for the first time, described the autopsy of a similar case in 1959, as follows: "the aortic valve was apparently normal, but the valve ring was separated from its attachment to the aorta." But further anatomical aspects could not be detailed in their report. We have found no other reports

of this condition. Recently, however, Hou et al.[6] reported a case of combined aortic and mitral regurgitation caused by a downward displacement of the left and noncoronary aortic annulus onto the anterior mitral leaflet. Although the possibility of superimposed rheumatic changes could not be ruled out, they considered its pathogenesis to be congenital.

There is no general agreement on the embryological pathogenesis of this condition. But it has been thought that, a semilunar valve develops from the bulbar ridges, which also takes part in the annular formation, and that the right coronary cusp originates from the ventricular myocardium[8]. Following point of view, the annular origin of the right aortic cusp, in our case, was excessively deep on the interventricular septum. Therefore, we emphasize the possibility of congenital abnormality to be the cause of aortic regurgitaion in this case. To our knowledge, our case is only the third report in the literature of a congenital aortic annulus displacement anomaly resulting in isolated aortic regurgitation.

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