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CASE REPORT

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Double-orifice mitral valve and an associated malformation: secundum atrial septal defect

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The authors report a case of double-orifice mitral valve (DOMV) which showed mitral stenosis and mild insufficiency. An associated anomaly was secundum atrial septal defect. DOMV is an unusual congenital heart defect. The occurrence of this anomaly with or without secundum atrial septal defect is very rare. More often it is associated with other congenital malformations arising from atrioventricular canal defects. There may be no haemodynamic consequences but mitral insufficiency and/or stenosis may complicate this malformation. Treatment can be summarised as abstention, surgical repair or valve replacement.

Key words: mitral valve, abnormalities, double-orifice, atrial septal defect

INTRODUCTION

Double-orifice mitral valve (DOMV) is an unusual but surgically important condition. It is often associated with atrioventricular (AV) canal defects, although it can be seen either as an isolated malformation or in association with other cardiac anomalies.

Double-orifice mitral value was first described by Greenfield in 1876 (guoted from [9]). Double-orifice or duplication of the mitral valve usually presents a congenital mitral stenosis or may show normal valve function [8]. Although this malformation of the valve usually produces mitral stenosis and, very rarely, mitral insufficiency; it may present no haemodynamic disturbance [5]. In half of the reported cases the duplication of the mitral valve orifice is seen as an isolated lesion [4].

We present here a case of DOMV which is supported by chordea tendinea originating from the papillary muscles and the AV wall. No mitral cleft or AV canal defect was associated with this valve, which showed severe mitral stenosis and mild insufficiency. A bridge of tissue separated both orifices, which were supported by leaflets that enabled the valve to be competent.

CASE REPORT

A 46-year-old female patient came to the hospital for the first time when she was 7 years old, complaining of effort dyspnoea and exercise intolerance. Echocardiography provided a diagnosis only of secundum type atrial septal defect (ASD). The secundum ASD was repaired primarily, no additional defect was detected during the operation and the patient was discharged with excellent postoperative progress. Three years later she was admitted again to Hacettepe University Department of Cardiovascular Surgery with dyspnoea on effort and palpitation. She was therefore diagnosed with congenital heart disease, probably mitral stenosis. In the light of this, she was given medical treatment. Her treatment over the following years was limited to the control of dys-

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pnoea. Then, in 1972, an echocardiographic examination revealed that the mitral valve might have a double orifice. However, the mitral stenosis was mild and the symptoms disappeared with medical treatment. She was followed up for almost 30 years with the focus on mitral stenosis. In 1994 the patient was admitted to hospital with refractory congestive heart failure. The diagnosis of DOMV was established during this admission. Moderate mitral stenosis and an insufficiency of 1–2° was detected in both mitral orifices on echocardiography. In the same year she gave birth normally at full term. Following her discharge from the hospital after delivery, the patient's cardiac status was regularly checked with echocardiography. In 2001 the mitral valve was replaced because of severe mitral stenosis and mitral insufficiency of 2°, diagnosed by colour-Doppler echocardiogram and left cardiac catheterisation. The peak diastolic gradient was 24 mm Hg (with a mean of 12 mm Hg). Cardiac catheterisation revealed a peak pulmonary capillary wedge pressure of 12 mm Hg and pulmonary artery systolic pressure of 35–40 mm Hg.

The patient underwent surgical correction. The two orifices of the mitral valve, which were supported by chordea tendinea originating from the papillary muscles and AV wall, were unequal in size. The larger (major) orifice was located posteriorly, and the other (accessory) orifice anteriorly. The two orifices were completely bridged by fibrous tissue (Fig. 1). The area was 1.34 cm^2 in the larger orifice and 0.4 cm^2 in the smaller. These results supported the echocardiographic findings. The valvular leaflets of the larger orifice had proper chordae tendineae and papillary muscles. Partially normal chordae tendineae originated from the anterolateral and posteromedial leaflets and were attached to the anterolateral-middle and posteromedial-middle papillary muscles respectively (Fig. 2). In addition, the mitral valve was highly fibrotic and also showed examples of severe stenosis and mild insufficiency. The excised valve was therefore replaced with St. Jude mechanical prosthesis No. 31 under cardiopulmonary bypass with moderate hypothermia (28°C).

DISCUSSION

Anatomical as well as haemodynamic observations have shown that double mitral orifices function as competent units. Advances in echocardiography have allowed more frequent detection of this entity by non-invasive techniques. Double-orifice mitral valve and additional lesions of each subdivision of the mitral valve were clearly shown by two-dimensional and colour-Doppler echocardiography by Segni et al. in 1986 [10]. In the present case DOMV was not diagnosed preoperatively with colour-Doppler echocardiogram until the 3rd admission. Because experience and techniques of echocardiography had not yet advanced sufficiently to show this anomaly with its anatomical details before 1980, most cases of DOMV in literature were diagnosed intra-operatively.

Double-orifice mitral valve may be of functional significance, causing mitral stenosis or incompetence and is usually associated with other cardiac anomalies [7, 11]. The one which most often accompanies it is endocardial cushion defect (partial persistent AV canal) [2]. Similarly, according to Rosenberg et al. [7], 25% of patients with DOMV have partial persistent AV canal and about 5% of patients with partial persistent AV canal have DOMV. This association suggests that DOMV and partial persistent AV canal have

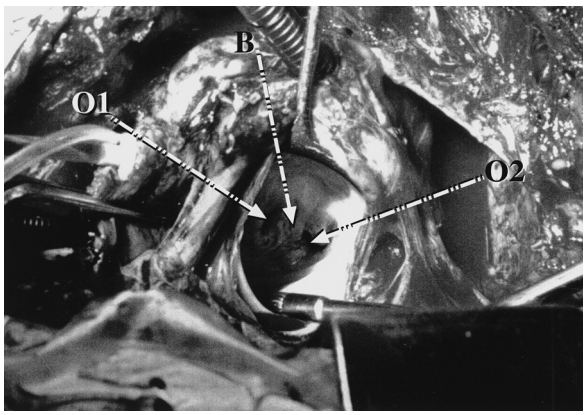

Figure 1. The appearance of the anomaly during the operation. O1 and O2 — indicate the larger and smaller orifices respectively, B — indicates the bridge between them.

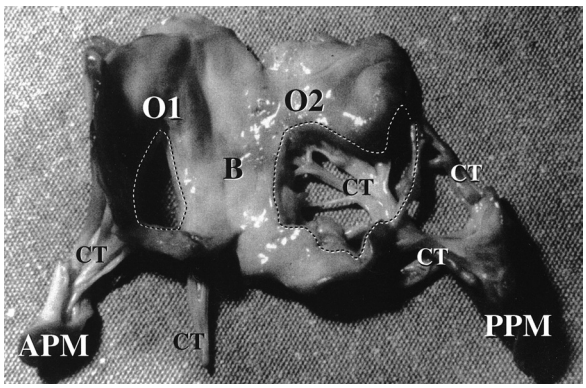

Figure 2. The appearance of the removed mitral valve. APM and PPM — indicate the anterior and posterior papillary muscles respectively, CT — indicates the chordea tendinea, O1 and O2 — indicate orifices, B indicates the bridge between them.

common developmental bases, such as a defect in the endocardial cushion [2]. However, in our case, there was only a secundum type atrial septal defect as an associated anomaly and this was closed in the first operation.

According to Baño-Rodrigo et al. [2], the frequency of equal-sized orifices in patients with DOMV is limited to 15%, with the remainder being unequal in size. In 48% of patients with double orifices the mitral valve function was normal, while mitral stenosis and regurgitation were seen in 26% each [9]. In our case, therefore, giving birth normally with such a malformation was extraordinary.

The framework that makes up DOMV can be divided into 3 forms: a complete bridge between the two orifices, an incomplete bridge, and a hole formed by the congenitally abnormal valves. A complete bridge (as in this case) is the structure that is least commonly seen [1].

Rosenberg et al. [7] stated that the true developmental basis of duplication of the mitral valve is unknown. DOMV might be the result of foetal endocarditis or may be a purely developmental anomaly [8]. In the only study so far which has put forward a logical hypothesis Lewis, after studying the Harvard collection of pig embryos, proposed that the double mitral valve was the result of an early arrest of development and that the accessory orifice represents a retention of the left portion of the common AV canal with subsequent reduction of the mitral ostium and alignment with it [2].

The non-cleft orifice of a DOMV is usually competent and rarely requires closure. The cleft, because it constitutes a type of parachute (single papillary muscle) valve, should be partially closed so as to relieve valve incompetence without causing undue stenosis. Repair of the AV canal associated with DOMV can be achieved with a low operative mortality and excellent late results [3]. Treatment may be summarised as abstention, surgical repair or valve replacement. It may cause peculiar surgical problems when plastic repair of the mitral valve is needed [6]. Metallic mitral valve replacement is preferable for fibrocalcific valves, mitral stenosis with insufficency, and also for giving birth to children and at a suitable patient age, as in our case [10].

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REFERENCES

1. Ancalmo N, Ochsner JL, Mills NL, King TD (1977) Double mitral valve. Report of a case and review of the literature. Angiology, 28: 95–100.
2. Baño-Rodrigo A, Praagh SV, Trowitzsch E, Praagh RV (1988) Double-orifice mitral valve: a study of 27 postmortem cases with developmental, diagnostic and surgical considerations. Am J Cardiol, 61: 152–160.
3. Domenico RD, Gheno G, Cucchini F (1993) Double orifice in prolapsing mitral valve. Inter J Cardiol, 41: 171–172.
4. Honnekeri ST, Tendolkar AG, Lokhandwala YY (1993) Double-orifice mitral and tricuspid valves in association with the Raghib complex. Ann Thorac Surg, 55: 1001–1002.
5. Lee CN, Danielson GK, Schaff HV, Mair DD (1985) Surgical treatment of double-orifice mitral valve in atrioventricular canal defects. J Thorac Cardiovasc Surg, 90: 700–705.
6. Lee DI, Ha JW, Chung B, Kim Y, Chun KJ, Rim SJ, Chung N (1999) Double-orifice mitral valve. Clin Cardiol, 22: 425.
7. Rosenberg J, Roberts WC (1968) Double-orifice mitral valve. Arch Path, 86: 77–80.
8. Sasaoka T, Ohguri H, Makita Y, Kurokawa S, Izumi T (1996) Double-orifice mitral valve in an elderly patient with tetralogy of Fallot. Jpn Heart J, 37: 503–507.
9. Saylam A, Oram A, Nazli N, Yener A, Aytaç A (1976) Double orifice mitral valve associated with ostium primum atrial septal defect. Turk J Pediatr, 18: 58–62.
10. Segni ED, Lew S, Shapira H, Kaplinsky E (1986) Double mitral valve orifice. Pediatr Cardiol, 6: 215–217.
11. Yurdakul Y, Arsan S, Karapinar K, Tamim M, Bilgiç A (1995) Congenital double-orifice mitral valve — a case report. Turk J Pediatr, 37: 173–176.