Severe mitral regurgitation in a woman with a double orifice mitral valve

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SUMMARY A 64 year old woman with partial atrioventricular canal and double orifice mitral valve presented with severe mitral regurgitation secondary to a torn leaflet. The double orifice regurgitant mitral valve is an unusual finding at operation.

Double orifice mitral valve is an unusual congenital abnormality that is rarely of clinical importance. We describe a patient in whom this finding was associated with severe mitral regurgitation.

Case report

A 64 year old woman was noted to have a murmur at age 20, but was symptom free until 1977 when congestive heart failure developed. A murmur typical of mitral regurgitation was noted. Her symptoms resolved on digoxin and diuretic treatment. She did well until November, 1983, when symptoms of weakness, abdominal distension, dyspnoea, and palpitation developed. Frusemide and quinidine (for ventricular extrasystoles) were started but when her symptoms progressed she was referred to our institution.

CLINICAL FINDINGS

Physical examination showed a normotensive female in no distress. Pulse was 66 beats per minute with frequent extrasystoles. Her venous pressure was raised with pronounced V waves. Carotid pulses were normal. The apical impulse was felt in the sixth intercostal space in the midclavicular line, and there was a left parasternal thrill. The first and second heart sounds were normal. The third heart sound was soft. A grade 5/6 harsh pansystolic murmur radiated from the heart apex to the axilla and spine. There was a grade 3/6 harsh systolic murmur at the left sternal border. There was a soft diastolic murmur at the apex. A pulsatile liver was palpable 5 cm

below the right costal margin. There was minimal oedema.

The electrocardiogram showed left axis deviation, first degree atrioventricular block, and right bundle branch block. A chest x ray film showed cardiomegaly, prominent pulmonary arteries, and increased pulmonary vascular flow. Laboratory studies were unremarkable.

Cross sectional echocardiography (Fig. 1) showed left atrial, right atrial, and right ventricular enlargement; an ostium primum type atrial septal defect; and a cleft anterior mitral leaflet. A contrast study demonstrated right to left shunting across the atrial septal defect. At retrospective examination of the echocardiogram two separate mitral valve orifices could be seen in the parasternal long axis right ventricular inflow view.

Cardiac catheterisation demonstrated raised right heart pressures, increased right atrial oxygen saturation and mild arterial desaturation. The pulmonary: systemic blood flow ratio was 1.9:1.

Left ventricular cineangiography showed normal wall motion with evidence of a gooseneck deformity of the left ventricular outflow tract and severe mitral regurgitation. There was left atrial and right ventricular enlargement. Early appearance of contrast in the right atrium confirmed the presence of an atrial septal defect. Coronary angiography was normal.

FINDINGS AT OPERATION

The heart was explored through a lateral right atrial incision. The tricuspid valve annulus was considerably dilated with a cleft in the septal leaflet extending to the septal rim. The valve was grossly incompetent. A 3 cm diameter ostium primum defect was present. The mitral valve was fully viewed through an incision in the atrial septum (Fig.

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Fig. 1 Cross sectional echocardiogram (parasternal long axis, right ventricular inflow view) showing both the atrial septal defect (ASD) and the dual orifice mitral valve. LA, left atrium; LV, left ventricle; MVO, mitral valve orifice; RA, right atrium; RV, right ventricle; TV, tricuspid valve.

2). It was composed of an oval annulus with two complete orifices divided by a transverse bridge of leaflet tissue. Each orifice had its own slightly eccentric placed papillary muscle. From each muscle, chordae tendineae radiated in a parachute fashion to the leaflet edges. The posterior orifice was approximately two thirds the diameter of the anterior orifice and was fully competent. The anterior orifice had a cleft in its anterior leaflet opposite the cleft in the septal leaflet of the tricuspid valve. On the anterior ostial margin of the cleft there was a single ruptured chord and a thickened flail leaflet. A regurgitant jet was demonstrated in this segment when the left

Fig. 2 Drawing of the in situ surgical findings. The mitral valve is viewed at operation through a surgically enlarged ostium primum atrial septal defect via a right atrial incision. Mitral valve (MV) with torn chord (c) and cleft anterior leaflet (large arrow) and bridging tissue (b). TV, tricuspid valve with cleft septal leaflet (small arrows). SVC, superior vena cava. IVC, inferior vena cava.

ventricle was filled with saline solution.

Repair of the ruptured chord by reimplantation did not restore valve competence. Cleft closure with interrupted sutures also failed to restore complete competence. We considered closure of the larger orifice, but believed that the secondary orifice would be inadequate to maintain flow. Therefore both mitral valve orifices were excised and were replaced with a 3M Starr-Edwards 6120 valve. The cleft in the septal leaflet of the tricuspid valve was closed. Tricuspid competency was restored by re-establishing a near normal annular shape and size by use of a 28 mm Carpentier tricuspid ring. The ostium primum defect was closed with a patch of autogenous pericardium secured by running suture.

Discussion

Double orifice mitral valve is a rare disorder that was first reported by Greenfield in 1876¹; over sixty cases have since been described.²⁻¹³ This abnormality is rarely of clinical importance,⁷ and most cases are recognised at operation or necropsy. It occurs as an isolated lesion or in association with other anomalies such as atrioventricular canal, bicuspid aortic valve, coarctation of the aorta, transposition of the great arteries, patent ductus arteriosus, supravulvar mitral stenosis, subaortic stenosis, secundum atrial septal defect, and ventricular septal defect.^{3 4 6 10 13} Endocardial cushion defects are present in 25-50% of cases,^{4 7 14} with double orifice mitral valve being seen in 3-10% of all cases of persistent atrioventricular canal.^{3 13 14} The case we describe is unusual because of the patient's age and mode of presentation. The oldest patient in the Wakai and Edwards necropsy study was 13 years old and all the others were <2½ years old.³ In the largest series reported all patients were ≤20, and the double orifice valve was generally an incidental finding.¹³ Clinically important mitral regurgitation^{5 6 9 15} has been diagnosed on the basis of a geometrically deformed valve,⁹ anterior mitral leaflet fenestration,⁵ or abnormal origin of the chordae tendineae and papillary muscles.⁶ This is the first report of a case in which a flail leaflet due to a ruptured chorda tendineae caused severe mitral regurgitation.

ANATOMICAL AND EMBRYOLOGICAL FEATURES

Ilbawi *et al* reported eight patients with double orifice mitral valves.¹⁴ Four had an intermediate atrioventricular canal without a cleft mitral leaflet while the others had ostium primum defects. The former group had intractable congestive heart failure and did not survive operation, whereas the latter

group recovered. These findings accord with those in a later series.^{3 13} All five of the cases in the Wakai and Edwards series were associated with complete or intermediate atrioventricular canals and all the patients died of congestive heart failure or post-operatively.³ The 11 cases in the Warnes and Somerville series were associated with ostium primum defects and tended to have a better course.¹³ This resembles our patient, who first presented in her fifties.

There is some disagreement about the embryological origin of this lesion. Wimsatt believed that the accessory orifice was the result of abnormal fusion between the anlagen of the medial and lateral mitral valve leaflets.¹⁶ Lewis, however, believed that the accessory orifice was the result of developmental arrest and persistence of the left part of the common atrioventricular canal, resulting in an accessory orifice in tissue destined to become a mitral leaflet.¹⁶ Ancalmo *et al* supported the latter theory because endocardial cushion defects are often found in association with this lesion.⁷

ANGIOGRAPHIC AND ECHOCARDIOGRAPHIC FINDINGS

Our patient showed the typical gooseneck deformity of the subaortic region of the left ventricle seen in ostium primum defects caused by abnormal placement of the anterior mitral leaflet.^{17 18} Warnes and Somerville and Somerville and Jefferson have described two rows of fine crenations in the medial region of the left ventricle, representing the double orifice.^{13 18} Echocardiography will often define both orifices.^{14 18} If an ostium primum defect is present, the fishmouth appearance of the normal mitral valve is replaced by a triangular configuration with separation of the anterior components of the anterior leaflet. Although the double orifice was not recognised before operation in our patient, on retrospective examination of the echocardiogram the angulated parasternal long axis view showed two orifices (Fig. 1).

SURGICAL CONSIDERATIONS

The surgical approach to patients with mitral valve abnormalities associated with endocardial cushion defects has recently been reviewed.^{14 19} Our approach resembles that of the Mayo Clinic group.²⁰ It has been our experience that when the cleft in the mitral valve has been left intact, or only partly closed, reoperation has been required because of progressive mitral regurgitation.¹⁹ Therefore, we usually close the cleft with interrupted sutures if the mural leaflet component of the mitral valve is of adequate size (that is, greater than one third of the circumference of the annulus). In cases where the

mural leaflet component is less developed, the cleft has been left partly or completely open; however, as noted earlier, this has given less than satisfactory results. In cases where the cleft is left open, invagination of the base of the commissure may provide better apposition and competence. If not, mitral valve replacement is probably indicated.

Double orifice mitral valve is a rare lesion that can complicate the surgical repair of endocardial cushion defects.^{14 18} Diagnosis should be considered in patients who have evidence of an ostium primum atrial septal defect. Cross sectional echocardiography and left ventricular angiography can be useful in establishing the diagnosis.

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