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CASE REPORT
Congenital heart disease

Congenital double mitral orifice with severe mitral regurgitation-associated rheumatoid arthritis: a case report

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Background A double orifice mitral valve (DOMV) represents a rare congenital malformation characterized by two valve orifices with two separate subvalvular apparatus. Double orifice mitral valve is congenital anomaly of the subvalvular mitral valve apparatus consisting of an accessory bridge of fibrous tissue, which partially or completely divides the mitral valve into two orifices.

Case summary A 30-year young male presented with dyspnoea and palpitation for 4 years, joint pain for 2 years and weakness of right upper limb and lower limb for 6 months. On clinical examination, Boutonniere, Swan neck, and Z-deformity of hand and foot metatarsal bone deformities are noted, on further evaluation, patient was diagnosed as a case of DOMV and was managed conservatively since patient was not willing for surgery.

Discussion Two-dimensional echocardiography is the best detection method, the parasternal short-axis view being most useful to show DOMV.

Keywords Double orifice mitral valve • 2D echocardiography • Mitral regurgitation • Rheumatoid arthritis • Case report

Learning points

- Congenital double mitral orifice is a very rare abnormality in which there are two separate orifices with separate respective chordal and papillary structures.
- Congenital double mitral orifice an accessory bridge of mitral valve tissue partially or completely divides the mitral valve into two orifices. Congenital double mitral orifice rarely occurs as an isolated anomaly but is most commonly associated with a variety of other cardiac anomalies of which atrioventricular septal defects are most frequent.
- Congenital double mitral orifice is usually detected at a young age but is an exceedingly rare diagnosis in adults. Echocardiography is a suitable modality for diagnosing congenital double mitral orifice and offers the possibility of comprehensive anatomical and functional assessment.
- Furthermore, this case reminds us that double orifice mitral valve is anatomically complex disease, it is commonly associated with other cardiac abnormalities and a precise anatomical and functional assessment is required. Our patient was advised surgical management but patient and his family did not give consent for surgery. He was managed conservatively and on follow-up, he improved symptomatically and has been on a regular follow-up for past 2 years.

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Introduction

A double orifice mitral valve (DOMV) represents a rare congenital malformation characterized by two valve orifices with two separate subvalvular apparatus. It is most often associated with atrioventricular septal defects but may also be present together with other congenital heart defects such as left-sided obstructive lesions, ventricular septal defects, or cyanotic lesions.[1,2] Rarely, patients with isolated DOMV are reported. However, even in the modern era of echocardiography DOMV often remains unrecognized. Double orifice mitral valve consists of two anatomically distinct mitral orifices that are divided by an accessory bridge of fibrous connective tissue. The fibrous ridge is composed of the mitral leaflet tissue and chordae tendineae. The embryogenic origin of DOMV is somewhat controversial. However, it is conceivable that the occurrence of abnormal differentiation of the mesenchymal endocardial cushion tissue into chordae instead of the leaflet tissue can result in such a defect. In DOMV, tensor apparatus is always abnormal and in the most common form, each orifice is separately attached to their individual, single papillary muscle, and this creates a kind of double parachute mitral valve,[3] as was observed in our case of DOMV, severe mitral regurgitation, and associated with rheumatoid arthritis.

Timeline

Four years prior to presentation	Dyspnoea and palpitation.
Two years prior to presentation	Joint pain.
Six months prior to presentation	Weakness of right upper limb and lower limb.
On presentation	Boutonniere, Swan neck, and Z-deformity of hand and foot metatarsal bone deformities are noted. Chest X-ray showed cardiomegaly with dilated left ventricle. Two-dimensional echocardiography showed congenital heart disease, SITUS solitus, double orifice mitral valve, and severe mitral regurgitation Grade III, mitral leaflets thickened, right mitral orifice 1.9 cm^2 left mitral orifice 2.4 cm^2.
After 10 days	A patient was managed conservatively since patient was not willing for surgery. The patient is on regular follow-up on medications for the past 2 years.

Case presentation

A 30-year young male presented with dyspnoea [New York Heart Association (NYHA) Class II] and palpitation for 4 years, joint pain for 2 years and weakness of right upper limb and lower limb for 6 months. He had no known medical history of diabetes, hypertension, rheumatic heart disease, or collagen vascular diseases. There was no other significant previous medical history. On examination, heart rate 80/min with high volume pulse, blood pressure 136/50 mmHg and had pansystolic murmur in the mitral area, no pulmonary crackles or rales. On clinical examination, Boutonniere, Swan neck, and Z-deformity of hand and foot metatarsal bone deformities were noted (*Figure 1A and B*). Routine blood investigations were normal, elevated erythrocyte sedimentation rate, and positive rheumatoid factor, VDRL, HIV, and complete haemogram were normal. Antinuclear antibody, anti-cyclic citrullinated peptide, C-reactive protein, and erythrocyte sedimentation rate were positive. Chest X-ray had evidence of cardiomegaly with dilated left ventricle. Electrocardiography revealed normal sinus rhythm. Two-dimensional echocardiography showed congenital heart disease, SITUS solitus, DOMV, and severe mitral regurgitation Grade III (*Figure 2A and B*), mitral leaflets thickened, right orifice 1.9 cm^2, left orifice 2.4 cm^2 (*Figure 2C*), and left ventricular internal diameter end-diastole 6.5 cm, left ventricular internal diameter end-systole 4.6 cm, left atrium 5.6 cm, end-diastolic volume 185 mL, end-systolic volume 65 mL, and left ventricular ejection fraction 65%. Three-dimensional echocardiography showed DOMV (*Figure 2D*). The patient was managed conservatively with angiotensin-converting enzyme inhibitors, ramipril 5 mg daily, lasilactone 50 mg, methotrexate 15 mg/weekly, bisoprolol 2.5 mg daily, since patient was not willing for surgery. He was managed conservatively and on subsequent follow-up patient improved symptomatically (with NYHA Class I dyspnoea). Complete blood count and liver function tests and echocardiography assessments were performed once in three months. On most recent follow-up, 3 months back transthoracic echocardiogram did not show any significant changes.

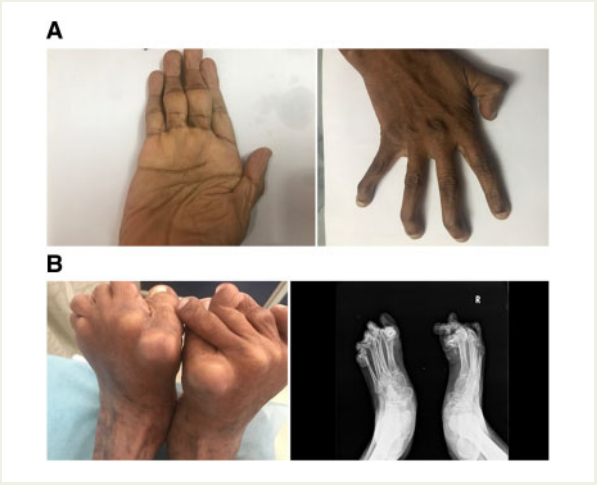

Figure 1 (*A*) Boutonniere, Swan neck, and Z-deformity of hand. (*B*) Deformity of metatarsal bone and X-ray foot showing metatarsal bone deformities.

We present a unique case of congenital double mitral orifice with severe mitral regurgitation associated rheumatoid arthritis. This case reminds us that DOMV is anatomically a complex disease, commonly associated with other cardiac abnormalities. In our case, severe mitral regurgitation associated with DOMV, surgical repair is a suitable

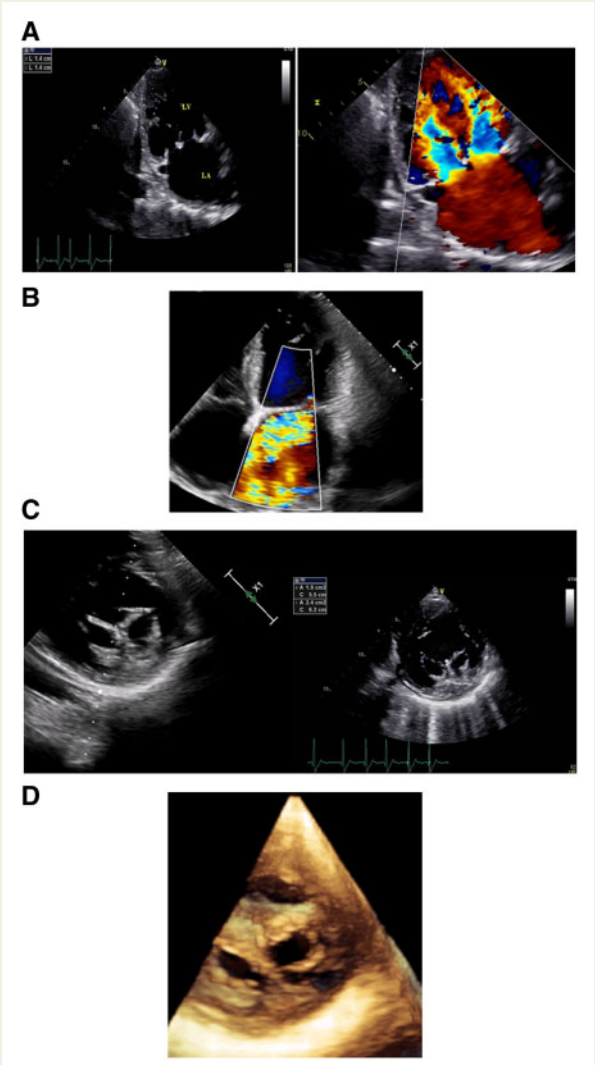

Figure 2 (*A*) Two-dimensional echocardiography two-chamber view showing double mitral orifice, and colour Doppler flow showing severe mitral regurgitation. (*B*) Two-dimensional echocardiography apical four-chamber view showing severe mitral regurgitation. (*C*) Two-dimensional echocardiography short-axis showing double mitral orifice with right orifice 1.9 cm^2 and left orifice 2.4 cm^2. (*D*) Three-dimensional echocardiography short-axis showing double mitral orifice.

treatment. As the patient is not willing for surgery, we have managed on medications and regular follow-up.

Discussion

Double orifice mitral valve is a very rare abnormality in which there are two separate orifices with separate respective chordal and papillary structures. This abnormality was first reported in 1876 by Greenfield, incidence of this rare abnormality to be 0.05%.[4] Exact incidence of DOMV is still unknown. However, in two retrospective studies on the evaluation of the echocardiographic findings of two referral centres, the incidence of DOMV has been found to be approximately 0.04% and 0.01% among children and adult population.[5] Double orifice mitral valve is found to be associated with transposition of the great arteries, ostium primum defect ostium secundum defect, and ventricular septal defect, atresia of the left ventricular outflow tract, single coronary artery, and as part of various syndromes with extracardiac and cardiac manifestations.[6] Patients with isolated DOMV are reported rarely.[7] Echocardiography is a suitable modality for diagnosing DOMV and offers the possibility of comprehensive anatomical and functional assessment.[8] Other concomitant congenital heart defects are detected during echocardiographic examination. Parasternal short-axis view in transthoracic echocardiogram is the best view for showing the numbers of mitral valve leaflets and orifices.[9] The management of DOMV is related to the type and severity of mitral valve dysfunction. Asymptomatic DOMV usually requires no active intervention. In case of severe mitral regurgitation or associated cardiac anomalies, surgical repair is necessary. A long-term follow-up is required for the early detection of complications of DOMV.[10] DOMV in association with rheumatoid arthritis is unreported. Based on the experience obtained in the present case, we emphasize the importance of meticulous transthoracic echocardiography examination to detect any valvular, structural malformations and defects in such cases to determine the best management plan.

Conclusion

This case demonstrates that careful examination is needed and if two separate mitral orifices are present that are supported by their own chordae from two different papillary muscles, then DOMV should be suspected. Furthermore, this case reminds us that DOMV is anatomically a complex disease, it is commonly associated with other cardiac abnormalities and a precise anatomical and functional assessment is required. Our patient was advised surgical management but patient and his family did not give consent for surgery. He was managed conservatively and on follow-up, he improved symptomatically and has been on a regular follow-up for the past 2 years.

Lead author biography

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Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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