Complicated rheumatic mitral stenosis presenting in an elderly patient and the challenges in its management

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SUMMARY

A 76-year-old lady with a recent diagnosis of rheumatic heart disease (RHD), and a history of repeated lower respiratory tract infections, came with symptoms of gastritis unrelated to the primary disease but further diagnostic study in the hospital revealed poorly controlled atrial fibrillation, grossly dilated left atrium with two large left atrial thrombi and mitral valve area $<1\text{ cm}^2$. It was decided that the best approach in our patient would be mitral valve replacement with mechanical prosthesis. Despite the usual trend of using bioprosthesis in the elderly, our decision was influenced by the fact that the patient would need chronic anticoagulation for atrial fibrillation in any case. The purpose of our case presentation is to illustrate a late-presenting case of RHD with unusual associations and the challenges to choose the best possible management.

BACKGROUND

Our patient, an unusually late-presenting case of rheumatic mitral valve disease with an incidental finding of massive left atrium, presented with numerous challenges in trying to decide the best possible treatment: old age, atrial fibrillation, requirement of long-term anticoagulation, left atrial thrombi and mitral valve calcification. By presenting this case, we aim to not only highlight an unusual case presentation of rheumatic heart disease but also provide a logical approach in deciding the treatment for similar cases with an emphasis on old age and long-term survival benefit. We also hope to emphasise the need to individualise the treatment, taking into consideration all the factors in a particular patient.

CASE PRESENTATION

A 76-year-old housewife from Mumbai came to the casualty department of Sir JJ Hospital with complaints of 5–6 episodes of vomiting since the morning that day. The vomitus was non-bilious and contained food particles. Her medical history was positive for repeated admissions for lower respiratory tract infection. Three years previously, she had been diagnosed with rheumatic mitral stenosis and atrial fibrillation following symptoms of occasional palpitations, dizziness and breathlessness. Echocardiography carried out at that time had shown critical mitral stenosis with left atrial enlargement. She had refused surgery earlier and the patient had since been maintained on ventricular rate control drugs. The patient did not remember having any symptoms suggestive of rheumatic fever in her childhood. She had no

history of stroke/transient ischaemic attack in the past.

On examination, she had irregular pulse at the rate of 108/min, blood pressure of 100/70 mm Hg, normal jugular venous pressure and a pale appearance. Chest examination revealed right-ventricular-type apical impulse on the left fifth intercostal space and a palpable parasternal heave and diastolic shock. Auscultation of the chest revealed first heart sound (S1) of variable intensity, loud P2, a diastolic murmur of variable length, no pre-systolic accentuation, faintly audible opening snap with variable A2-OS interval and a systolic murmur in the tricuspid area which increased on inspiration.

INVESTIGATIONS

ECG showed right bundle branch block and atrial fibrillation with a ventricular rate of 110/min (figures 1 and 2). Chest roentgenogram (figure 3) showed hugely dilated left atrium, right atrium and right ventricle with double density sign and convex left atrial appendage. CT of the chest had been carried out a month back, when she presented with chest pain, cough and expectoration. It was carried out to rule out pulmonary embolism and for the evaluation of possible lower respiratory tract infection (LRTI). It incidentally revealed a massively dilated left atrium ($11\times 10\text{ cm}$), right atrium ($9.5\times 8\text{ cm}$) and right ventricle with reflux of blood seen in the inferior vena cava and hepatic veins. Mitral valve showed calcification. It also revealed two filling defects/thrombi, one attached to the anterior wall of left atrium of size $6.5\times 3.7\text{ cm}$ and the second attached to the posterior wall of size $3.2\times 2.1\text{ cm}$ (figures 4–6). Results of two-dimensional echocardiography included thickening of mitral valve, mitral valve commissural calcification with a valve area of 0.9 cm^2 on planimetry, an echo score of 8/16 and confirmed the presence of two thrombi in the left atrium. M-mode echocardiography showed reduced left-ventricular internal dimensions at diastole and systole, reduced-end systolic and diastolic volumes, a normal ejection fraction (60.2%) and left atrial dimension of $8.6\text{ cm}\times 7.8\text{ cm}$. Colour doppler showed a mitral valve area of 0.9 cm^2 , estimated pulmonary artery systolic pressure of 70 mm Hg (normal: 15–30 mm Hg) and grade 1 mitral regurgitation. Coronary angiography ruled out any significant coronary artery disease.

DIFFERENTIAL DIAGNOSIS IF RELEVANT

She was diagnosed as a case of gastritis in a known case of rheumatic heart disease and mitral stenosis

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Figure 1 ECG of the patient at presentation showing atrial fibrillation with rapid ventricular rate, right axis deviation, RBBB and no evidence of P waves.

Figure 2 ECG of the patient, taken 10 days later, showing better ventricular rate control on β -blockers.

complicated by pulmonary hypertension, atrial fibrillation and left atrial thrombi. The gastritis seemed to be unrelated to the underlying rheumatic heart disease, but the intensive diagnostic study drew attention to the primary problem of severe mitral stenosis.

TREATMENT

The patient had previously refused surgery and she had been maintained on ventricular rate control drugs. During the recent presentation, she was again explained the procedure, the alternatives and the expected outcome and she agreed to undergo surgery in view of the long-term risks. She subsequently underwent mitral valve replacement with mechanical prosthesis and thrombectomy. She was also started on anticoagulation with warfarin with a target international normalised ratio (INR) range of 2.0–3.0.

OUTCOME AND FOLLOW-UP

The patient showed good recovery in the immediate post-operative period and regular follow-up and, subsequently, showed a good response to the surgery and better ventricular rate control with drugs. There was a gradual fall in pulmonary artery systolic pressure following surgery.

DISCUSSION

Rheumatic heart disease has been the cause of significant morbidity and mortality in developing countries. An autoimmune disorder, it has been known to predominantly affect younger individuals following a throat infection with group A streptococci. Mitral stenosis, associated with rheumatic heart disease, can remain asymptomatic or even present as late as middle age or beyond. In such cases, the patient may not always remember about an attack of rheumatic fever in childhood, giving the

impression of late onset rheumatic heart disease. We report a case of an elderly woman who was recently diagnosed with rheumatic heart disease and mitral stenosis. Although it is difficult to comment when the disease actually started in our patient, we believe that the heart disease had been benign

Figure 3 Chest roentgenogram showing massive cardiomegaly and gross dilation of left atrium with double density sign (black arrows) and convex left atrial appendage (white arrow).

Figure 4 CT of thorax (sagittal section) showing enlarged left atrium and two attached thrombi on anterior and posterior walls.

Figure 5 CT of thorax (coronal section) showing the thrombus on the anterior wall.

until its presentation at an age well beyond the usual age of presentation in rheumatic heart disease. By this case report, we also try to discuss the possible therapeutic approaches to an elderly patient with critical mitral valve disease with significant calcification.

Massive enlargement of left atrium in patients with rheumatic heart disease has been reported before. Although mitral regurgitation has been postulated to profoundly influence this

association,¹ it is not uncommon to find it in patients with mitral stenosis alone or in predominance.² Our patient with giant left atrium was found to have dominant mitral stenosis and only mild-grade mitral regurgitation. Park *et al*³ reported a case of giant left atrium with critical mitral stenosis but only mild mitral regurgitation. Okyay *et al*⁴ reported a massively enlarged left atrium with a similar finding. The asymptomatic massive enlargement of left atrium (LA) in our patient may be

Figure 6 CT of thorax (cross-section) showing the thrombus on the posterior wall.

A/C = anticoagulation; AVR = aortic valve replacement; INR = international normalized ratio; MVR = mitral valve replacement

Another question was the choice of prosthesis: mechanical or biological. In patients >70 years of age, biological valves are preferred.⁹ This is predominantly because of lower life expectancy in these patients, non-requirement of anticoagulation with these type of valves and lower rates of bioprosthetic deterioration in the elderly.⁹ ¹⁰ However, a review of the literature raised some interesting points. Sidhu P *et al*¹¹ performed a comparison study over the long-term efficacy of mechanical and bioprosthesis over a period of 20 years in patients over the age of 70. The study concluded that bioprosthesis does not provide any survival benefit over mechanical valves in the elderly. Mechanical prosthesis has been known to be more durable, but this advantage is

Before undergoing surgery, the need to conduct a proper diagnostic study for comorbidities, including coronary angiography, could not be overemphasised. Coronary artery disease is common in this age group and may contribute to intraoperative mortality.

- ▶ The presentation of mitral stenosis may be delayed as the valvular disorder may follow a benign course compatible with normal life. It is not uncommon for mitral stenosis to present as late as old age.
- ▶ Asymptomatic massive enlargement of LA may be related to high LA compliance and a slow course of the valvular disease.
- ▶ Anticoagulation in the elderly with atrial fibrillation needs to be used with caution. Proper risk stratification and monitoring are required and the lowest possible dose of anticoagulation needs to be used.
- ▶ Balloon mitral valvuloplasty may not be an effective option in patients with significant commissural calcification and low echo scores.
- ▶ The choice of prosthesis should be determined on an individual basis, considering the presence of co-morbidities and other contributory factors like need for anticoagulation.

Patient consent Obtained.

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