Case Report

Isolated dextrocardia and rheumatoid arthritis with mitral stenosis with pulmonary hypertension with atrial fibrillation: A rare association

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ABSTRACT

This case presents a rare occurrence of rheumatoid arthritis (RA) in a 50-year-old woman with concurrent mitral valvular disease and dextrocardia. Diagnosed with seropositive RA 5 years earlier, the patient exhibited fever, cough, and progressive dyspnea. Clinical examination revealed irregular tachycardia, elevated jugular venous pressure, and signs of heart failure. Laboratory results confirmed seropositive RA, anemia, and elevated inflammatory markers. Electrocardiogram and echocardiogram indicated dextrocardia, atrial fibrillation, and moderate mitral stenosis. This case highlights the unusual association of RA with valvular heart disease and the added complexity of dextrocardia. Recognizing diverse cardiac manifestations in RA is crucial, contributing to the growing evidence linking autoimmune disorders to cardiovascular complications. Further research is essential to understand the intricate relationship between RA and unique cardiac abnormalities.

Key words: Atrial fibrillation, Dextrocardia, Rheumatoid arthritis, Valvular heart disease

Rheumatoid arthritis (RA) is the most common autoimmune disease with a global prevalence of 460 per one lakh population between 1980 and 2019 and occurs more frequently in women [1]. The valvular lesions caused by RA have been known for a long time. The valves may be affected by unspecific inflammatory changes or specifically by rheumatoid granulomas identical with subcutaneous nodules that lead to the malfunctioning of the aortic or the mitral valve.

Hereby, we present a patient affected by classical seropositive RA accompanied by mitral valvular disease with dextrocardia. There have been some reports of cases of dextrocardia associated with RA (rheumatoid heart disease) in which the heart was primarily affected at the mitral (tricuspid) valve and the aortic valve.

CASE REPORT

A 50-year-old female patient who was diagnosed with RA 5 years ago presented to us with a history of fever, productive cough, breathlessness, and palpitations for 1 week. There was no history of chest pain and syncope. For the past 3 years, the patient had breathlessness which was insidious in onset and slowly progressed from the New York Heart Association Grade 1 to Grade 4. This was associated with palpitations of an irregular nature which were not associated with chest pain, syncopal attacks, or post-palpitation diuresis.

On examination, the patient was dyspneic at rest with a pulse rate of 124 beats/min, which was irregularly irregular, and an apex pulse deficit of 24 beats/min. The patient had an elevated jugular venous pressure of 10 cm with an absent "a" wave and prominent "V" wave with "Y" descent. The patient also had bilateral pitting pedal edema, mild pallor, and central cyanosis. Cardiovascular examination revealed apical impulse being situated on the right side in the fifth intercostal space 1 cm lateral to midclavicular line. The patient had tachycardia with varying intensity of first heart sound with a mid-diastolic murmur and tricuspid regurgitation murmur. On examination of the respiratory system, the patient had tachypnea with bilateral fine crepitation in the infra-scapular and infra-axillary areas. Per-abdominal examination revealed a pulsatile liver.

Routine hematological investigations revealed normocytic hypochromic anemia with a hemoglobin of 9.4 g% and elevated erythrocyte sedimentation rate (57 mm/h). Immunological studies confirmed seropositive RA (RA factor: 151 IU/mL) and anti-ds DNA antibody (5.9 IU/mL). Antinuclear antibody test (ELISA) was negative. Biochemical investigations were within normal

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limits. Electrocardiogram showed evidence of dextrocardia with atrial fibrillation, and chest X-ray showed features of dextrocardia with left lower lobe pneumonia (Figs. 1 and 2). Echocardiography revealed situs solitus, dextrocardia with moderate mitral stenosis (MVO 1.45 cm^2) with severe tricuspid regurgitation and moderate pulmonary hypertension with nodular thickening of the mitral valve (Fig. 3).

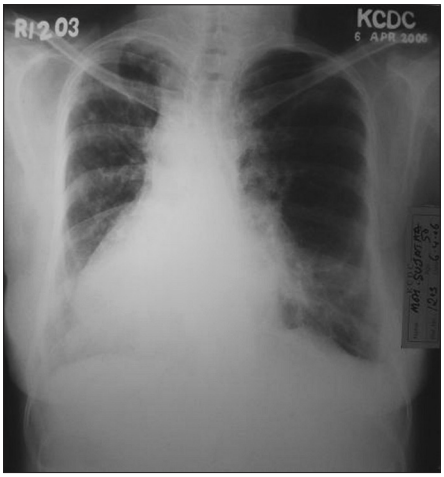

Figure 1: Chest X-ray posteroanterior view showing dextrocardia with situs solitus with left lower lobe pneumonia

DISCUSSION

RA is a common chronic autoimmune disease involving many organ systems, primarily the joints; frequently, it is accompanied by cardiac lesions including pericardium, myocardium, and endocardium, comprising coronary arteries, valvular tissue, and conduction system. It reduces both quality of life and life expectancy. Dextrocardia is mainly a congenital heart abnormality in which the heart is displaced to the right (dextroversion). Just as with levocardia, the heart can be atrial inversion with situs inversus or ambiguous. The most common variant is dextrocardia (situs inversus) with approximately 10% of cases being associated with additional congenital heart anomalies and about 25% are associated with Kartagener syndrome, which is related to sinusitis (sinusitis) and bronchiectasis [2].

Heart involvement in RA is the leading cause of death [3,4] Cardiovascular is the leading cause of death in patients with RA. Cardiac lesion causes 40–50% of mortality in RA patients [4]. Although the association between chronic arthritis and heart disease has been recognized for nearly 200 years, it was only in 1944 that Baggenstoss and Rosenberg described the presence of typical rheumatoid granuloma in the heart [5]. Waters discussed the concept of rheumatoid heart disease as a distinct pathological

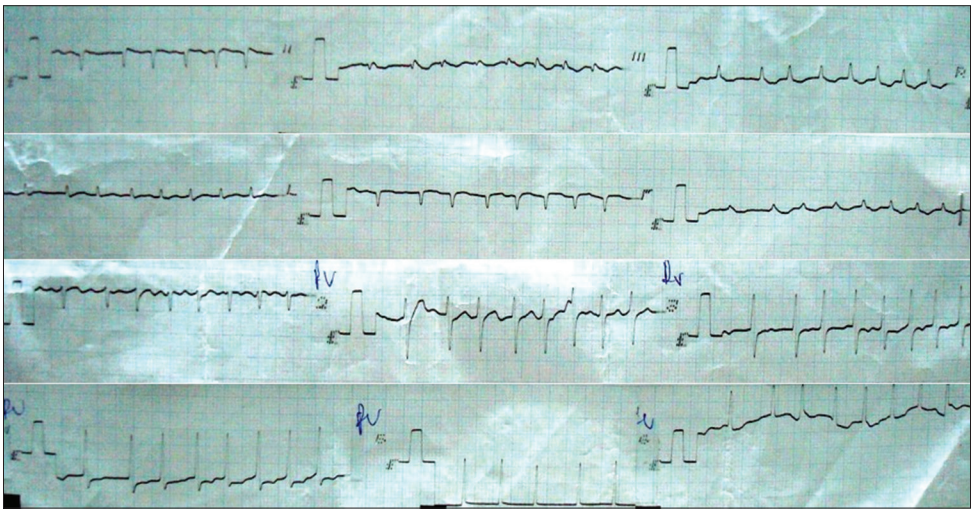

Figure 2: Electrocardiogram with right-sided lead showing dextrocardia with atrial fibrillation with right-sided chest leads

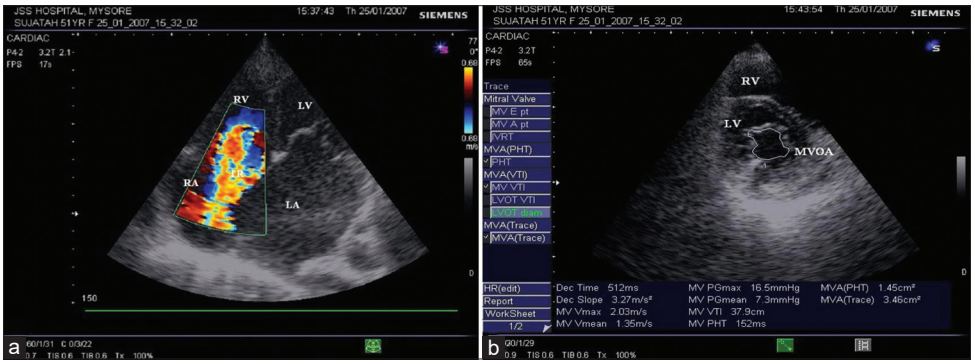

Figure 3: (a) Echocardiography with color Doppler (four chamber view) showing tricuspid regurgitation with diastolic doming of the anterior and posterior mitral leaflet with nodular thickening of the valve (rheumatoid nodule); (b) Echocardiography (apical view) showing nodular thickening of the mitral valve with an orifice area of 1.45 cm^2 with pulmonary hypertension

entity and this was confirmed by subsequent necropsy and clinical studies. After that, cardiac involvement in RA has been extensively described with variable results according to whether the lesion was sought by clinical examination, autopsy, or echocardiography.

Although pericarditis has been the most commonly reported abnormality, conduction disturbance and lesions of the valve, myocardium, and coronary arteries have also been described. RA with valvular heart disease (VHD) has not been well characterized and its clinical predictors are not defined [6]. The incidence of rheumatoid valvular disease is between 2.5% and 30% of necropsy patients with the above disease [6]. Diffuse or localized valve nodules and valve thickening by unspecific inflammatory changes were found in the aortic and mitral valves [7,8]. There was no correlation in RA between valvular disease and duration, activity, severity, pattern of onset and course, extra-articular disease, serology, or therapy [6].

Roldan *et al*., found mainly left VHD, in which valve nodules were more common and valve stenosis was found only in 1% of cases (aortic stenosis and mitral stenosis) [6]. Guedes *et al*., showed that cardiac involvement particularly the mitral valve is extremely common in RA [4]. Very few cases of association of Kartagener syndrome with RA are reported worldwide. However, in our case, the patient had isolated dextrocardia with RA along with mitral stenosis with atrial fibrillation. We hypothesize that the mitral valve changes have occurred due to RA as the patient had no history of rheumatic heart disease. The nodular thickening of the mitral valve is suggestive of RA. The cardiorespiratory symptoms started after 3 years of the onset of joint pain which also supports the etiological diagnosis of RA. There have been some reports of cases of dextro-heart disease associated with RA (rheumatoid heart disease) in which the heart was primarily affected by the mitral (tricuspid) valve and the aortic valve [9]. The dextro-heart disease is very rare, but the clinical presentation and treatment are like that of levo cardiography and angina pectoris [9,10]. In our case, the association of RA with VHD is rare but a known entity, however, dextrocardia is extremely rare. Further studies are needed to analyze these rare associations.

CONCLUSION

This case report highlights a rare presentation of RA with dextrocardia and mitral valvular disease. Understanding the diverse cardiac manifestations of RA is crucial for comprehensive patient management and underscores the need for further research to elucidate the complex interplay between RA and cardiovascular complications.

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