

A giant fibrinoid pericardial mass in a patient with rheumatoid arthritis: a case report

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Background

Rheumatoid arthritis (RA) is a chronic inflammatory disease of the joints, which may extend to extra-articular organs. Extra-articular manifestations have been considered as prognostic features in RA, and pericardial disease is one of the most frequent occurrences. Rheumatoid arthritis pericarditis is usually asymptomatic and is frequently found on echocardiography as pericardial thickening with or without mild effusion. Severe and symptomatic cases are rare, but pericardial masses are even rarer. We report a patient with erosive, nodular seropositive RA, and progressive functional deterioration owing to a giant pericardial mass compressing the right cardiac chambers.

Case summary

The patient was a 79-year-old man. Cardiac magnetic resonance imaging revealed a pericardial lesion measuring $10 \times 9 \times 6$ cm with complex structures in its interior, which had compressive effects on the right atrium and right ventricle, severely limiting diastole. Late gadolinium enhancement of the lesion walls and pericardium suggested pericarditis. Surgical resection was performed, and a soft mass with liquid content was extracted. The patient recovered well with improvements in symptoms and the functional status. Histopathological studies ruled out neoplasm, vasculitis, and infection, and the entire mass showed fibrinoid material associated with fibrinoid pericarditis.

Discussion

Symptomatic RA pericarditis is a rare cardiac manifestation of RA, whilst associated significant haemodynamic compromise is even rarer. The condition could manifest with a giant compressive pericardial mass composed of fibrinous material, with particular involvement of the right ventricle. Exclusion of other conditions, such as neoplasms and infections, is necessary.

Keywords

Arthritis • Rheumatoid • Pericardial effusion • Rheumatoid nodule • Heart failure • Case report

Learning points

- Symptomatic pericarditis due to rheumatoid arthritis is rare and can be related to a giant fibrinoid pericardial mass compressing the right cardiac chambers.
- In this case, a multimodality imaging approach determined the nature of the pericardial mass and extent of haemodynamic significance and aided surgical planning.
- Histopathological analysis ruled out chronic lymphoid infiltrates, vasculitis, granuloma, or neoplasia, and the entire mass showed fibrinoid material associated with fibrinoid pericarditis.

Introduction

Subclinical pericardial compromise is the most common cardiac issue in patients with rheumatoid arthritis (RA). Asymptomatic pericardial thickening with or without effusion can be noted on echocardiography in 30–50% of RA patients.¹ Only 2–4% of patients have symptoms, and <0.5% of patients experience cardiac tamponade or constrictive pericarditis.² In the cases of a clinically significant pericardial disease, multimodality imaging provides anatomical, physiological, and structural information that can help to achieve a precise diagnosis

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and optimal management.³ The differential diagnosis of RA pericarditis includes tuberculosis, cancer, other autoimmune diseases, and hypothyroidism, with different prognostic and therapeutic implications.⁴ On the contrary, pericardial masses are rare. Primary cardiac tumours are relatively rare, with a 0.001–0.03% incidence among autopsy cases, and they have a broad differential diagnosis.⁵ Pericardial tumours are less common than other cardiac tumours, and they account for only 12% of all primary cardiac tumours.⁶ Thus, multimodality imaging is considered highly valuable in the evaluation and treatment planning of pericardial masses.

Here, we report a case of an RA patient with heart failure due to a giant loculated mass of fibrinoid material compressing the right cardiac chambers, which was related to RA symptomatic pericarditis and was evaluated through multimodality imaging.

Timeline

| | |
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| Day 1 | A 79-year-old patient with active and erosive rheumatoid arthritis presented with acutely decompensated heart failure with predominant right heart congestion signs. Computed tomography indicated a giant mass at the pericardium, and the patient was admitted for further assessment and treatment |
| Day 2 | Echocardiography showed preserved left ventricle function but revealed a hyperechogenic mass compressing right heart chambers with a dilated Inferior vena cava. A cardiac magnetic resonance (CMR) was indicated. |
| Day 4 | Control of symptoms and congestion was achieved with medical management of heart failure. |
| Day 5 | CMR imaging demonstrated a pericardial lesion measuring $10 \times 9 \times 6$ cm with complex structures in its interior, which had compressive effects on the right atrium and right ventricle, severely limiting diastole. Late gadolinium enhancement of the lesion walls and pericardium suggested pericarditis. |
| Day 6 | This case was discussed by the cardio-surgical board (heart team), and surgical excision and pericardial decompression were recommended. Consent was obtained for the procedure. |
| Day 8 | Intraoperatively, severe thickening of the entire pericardium was noted. Additionally, a large soft mass with zones and liquid content was identified. The mass firmly adhered to the right atrium, superior and inferior vena cava, and right ventricle. Complete resection of the mass and anterior pericardiectomy was performed. |
| Day 13 | In macro and microscopic analyses, no area with chronic lymphoid infiltrates, vasculitis, granuloma, or neoplasia was observed. The entire mass showed fibrinoid material. The histopathological diagnosis corresponded to fibrinoid pericarditis. |
| Day 18 | The patient was discharged without major complications and with New York Heart Association functional Class II. The patient underwent a cardiac rehabilitation programme and received rheumatology and cardiology services. |

Case presentation

A 79-year-old man presented to the emergency department with progressive dyspnoea, orthopnoea, an augmented abdominal perimeter, lower extremity oedema, and weight gain. He had a history of seropositive RA for over 10 years, which manifested as erosive polyarticular arthritis with rheumatoid nodules on the skin. He was being treated with prednisolone (5 mg daily), chloroquine (250 mg daily), sulfasalazine (500 mg three times a day), and leflunomide (20 mg daily) for at least 5 years. Additionally, he was being treated for heart failure involving a preserved ejection fraction (60%) with furosemide (40 mg daily), spironolactone (25 mg daily), and digitalis/drops (six times daily; 0.6 mg/mL) for the last 5 years. Moreover, he was being treated with metformin (850 mg) at his two main meals for diabetes. Furthermore, he was being treated with levothyroxine (125 µg daily) for hypothyroidism. He was currently a heavy smoker.

On examination, he appeared chronically ill and pale, with no signs of respiratory distress. His heart rate (76 b.p.m.), blood pressure (100/60 mmHg), and respiratory rate (16 breaths/min) were normal. He had a raised jugular venous pressure at 45°. His cardiac auscultation was normal; however, pulmonary auscultation revealed bi-basal hypoventilation. Additionally, an abdominal examination revealed ascites, collateral circulation, hepatomegaly, and lower extremity Grade III oedema. Multiple nodules that appeared hard, painless, and mobile were found on his hands, elbows, knees, lower back, and buttocks. There was tenderness with palpation and passive movement pain in his hands, wrists, shoulders, and elbows associated with movement limitation.

Administration of an intra-venous diuretic was initiated, and a positive clinical response was noted. Chest radiography revealed pleural thickening and fibrotic tracts in the left inferior lung base (*Figure 1A* and *B*). A thoracic computed tomography (CT) scan performed 10 months before the admission showed a single 10×11 -mm nodule at the anterior segment of the right superior lobule, pleural thickening with fibrotic tracts, and a low-density area of 8×6 cm with clear edges in contact with the right heart chambers (*Figure 1C*). Since then, the patient lost to follow-up. Transthoracic echocardiography (TTE) showed normal dimensions and functions of the left chambers, with a left ventricular ejection fraction of 50–55%. No significant valvular disease was noted. The estimated pulmonary systolic artery pressure was 30 mmHg. Additionally, a mass was noted over the right chambers, which severely restricted diastole (*Figure 1D* and *E*). Although there was no E-wave variability during breathing, the inferior vena cava was dilated without collapsibility, raising the possibility of a physiology similar to cardiac tamponade. The N-terminal pro-brain natriuretic peptide level was 347 pg/dL (local upper limit of normal, 125 pg/mL).

Cardiac magnetic resonance (CMR) imaging revealed a well-limited intra-pericardial multiloculated mass located anterior to the right chambers (*Figure 1F* and *G*). There was flattening of the interventricular septum without variability during the respiratory cycle, making a constrictive physiology unlikely, but the limitation of right ventricle diastole was prominent (*Supplementary material online, Video S1*). The mass appeared hyperintense in T2-weighted images and hypo-intense in unenhanced T1-weighted images, with a thick hyper-enhancing capsule without any intralesional solid or nodular components and no septal contrast uptake (*Figure 1H* and *I*). These

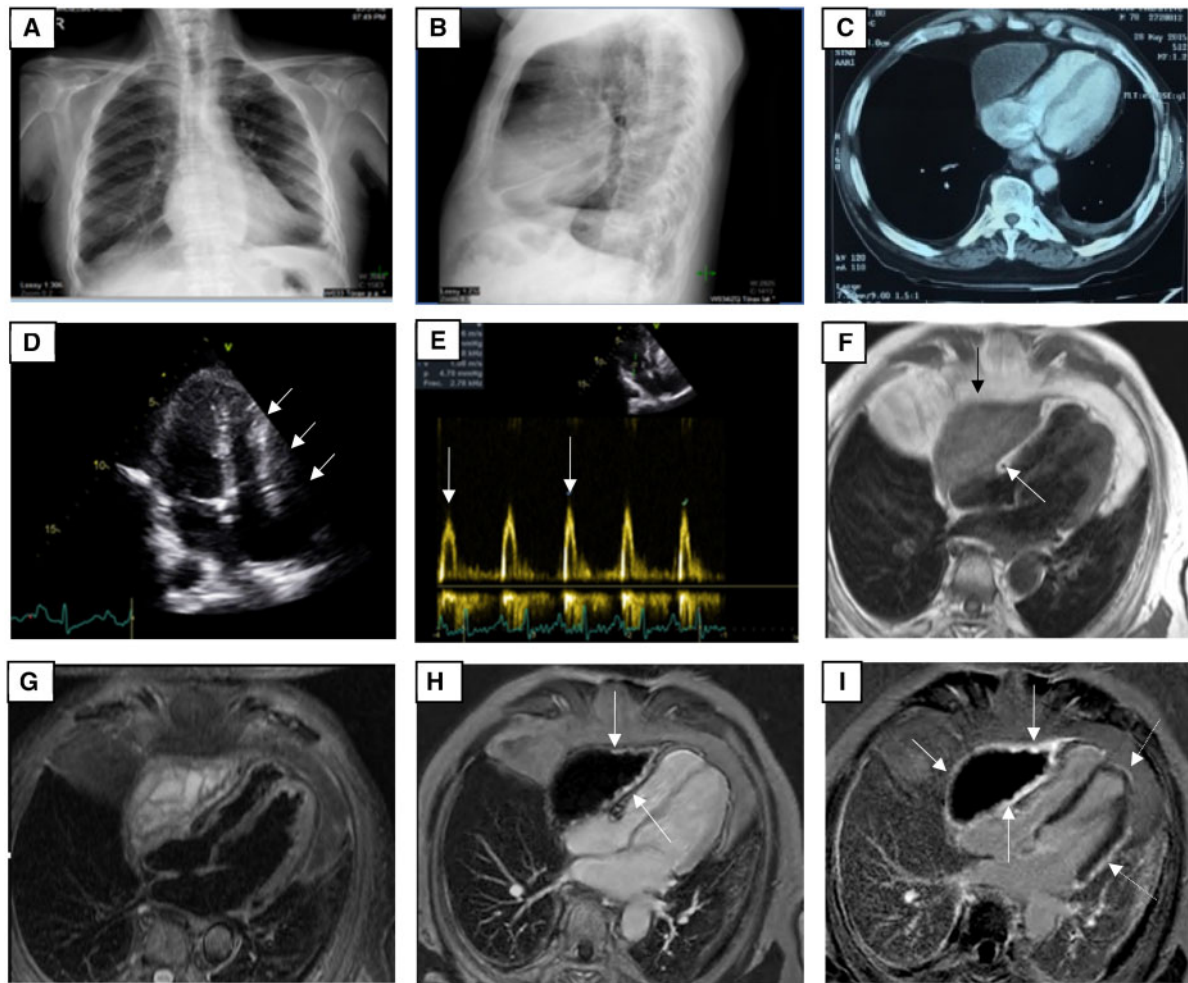


Figure 1 (A and B) Chest radiograph showing pleural thickening and fibrotic tracts in the left inferior lung base. (C) Computed tomography image showing a low-density area with well-defined limits in contact with the right heart chambers. (D) Transthoracic echocardiography image in apical four-chamber view with the left chambers on the left showing a hyperechoic structure that comprises the free wall of the right ventricle (white arrows). (E) No E-wave variability is seen during normal breathing (white arrows, inspiration and expiration). (F) Cardiac magnetic resonance T1 turbo-spin echo image in the four-chamber long-axis view showing a lenticular pericardial mass compressing the right ventricle free wall, right atrio-ventricular groove, and right atrium, with well-defined borders, sparing the epicardial fat. It has a mild hyperintense signal when compared with the adjacent myocardium (the black arrow indicates the mass; the white arrow indicates the right coronary artery). (G) Four-chamber T2 with fat suppression image showing that the mass has a fluid intensity signal. Several thick septae and thick capsules are noted, without free pericardial fluid or a hyperintense myocardial signal. (H) Four-chamber inversion recovery early gadolinium image showing rim enhancement (white arrow) without any intralésional contrast uptake. There are no intra-cavitary filling defects. (I) Four-chamber inversion recovery late gadolinium image clearly showing thick rim enhancement without any internal contrast uptake, which is consistent with inflammatory intra-pericardial fluid collection (white arrows). There is remote pericardial enhancement (white arrows with a dotted line) without myocardial enhancement, which is compatible with pericarditis without myocarditis.

findings raised the possibility of complex fluid collection associated with an inflammatory pericardial process. The lesion measured $10 \times 9 \times 6$ cm, and there was no mass infiltration or free pericardial effusion.

There was also global pericardial enhancement with mild thickening of the pericardium compatible with pericarditis (4 mm). Additionally, thickened and enhanced pleurae were seen on both sides.

Surgical excision of the mass with pericardial decompression was advised, and patient's consent was obtained. Intraoperatively, severe

thickening of the entire pericardium and a large soft mass were noted. Complete resection of the mass was performed with right and left anterior pericardiectomy. In macroscopic histopathological analysis, the mass appeared multiloculated with solid yellow pseudo-laminated areas that were mixed with irregular dark areas of less consistency. The entire mass showed fibrinoid material (Figure 2). Pericardial samples showed chronic multifocal inflammation and areas with organized fibrinous exudate. There were no areas with chronic lymphoid infiltrates, vasculitis, granulomas, or neoplasia. The

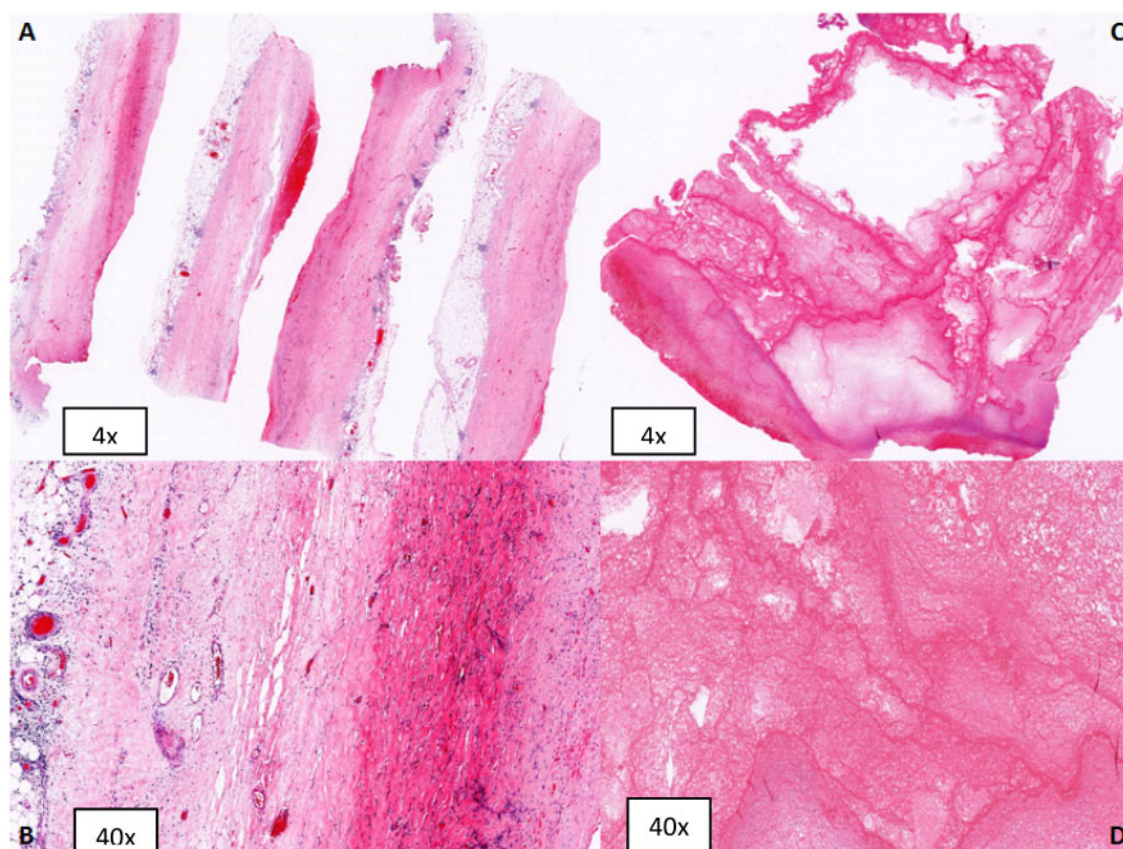


Figure 2 (A and B) Haematoxylin and eosin staining images of three pericardium slices showing collagenisation and thickening, with presence of mature lymphoid infiltrate into the outer zone with extension to the adipose tissue (arrow). No chronic granulomatous inflammation, acute inflammation, microorganisms, or malignant neoplasia. (C and D) Microscopic appearance of fibrinoid pericarditis that was in contact with pericardial wall.

histopathological diagnosis was consistent with fibrinoid pericarditis. The patient recovered well and was discharged 18 days after admission without complications and with notable improvements in symptoms and New York Heart Association functional Class II.

Discussion

We reported an RA patient with a large pericardial mass. A pericardial mass associated with RA is rare. To date, only three such cases have been reported in the medical literature.⁷⁻⁹ These cases were reported in 2006, 2012, and 2015 and involved three men aged 75, 69, and 65 years, respectively. The patients were admitted owing to fatigue and dyspnoea. All had longstanding poorly controlled seropositive RA and had a large pericardial mass that compressed the right heart structures. All the patients required surgical excision, and the surgical pathology indicated fibrinoid pericarditis.

Although new imaging modalities, apart from conventional TTE, are being used to diagnose pericardial diseases,¹⁰ TTE has an essential role in the diagnosis of pericardial pathology, especially due to its ability to accurately estimate haemodynamic compromise. Detailed anatomical characterization of the pericardium with cardiac CT and CMR imaging has facilitated the study of the aetiology of pericardial

diseases. Cardiac magnetic resonance imaging has the highest sensitivity and specificity for the detection of pericardial thickening and inflammation through evidence of oedema and contrast enhancement.^{11,12} A mass-like lesion is best evaluated using CMR imaging to differentiate among pericardial thickening, effusion, and infiltrative diseases, such as neoplasm.^{13,14} Real-time cine cardiac CT and CMR imaging can provide information similar to that with TTE concerning interventricular septal motion and chamber collapse.³ Nevertheless, there can be false-negative haemodynamic evaluation or incomplete criteria of constriction when the filling pressures are altered owing to marked diuresis, as it appears to have occurred in our case.³

In this case, CMR suggested a benign and inflammatory nature that was confirmed in the histological analysis. The RA pericardial compromise is characterized by the presence of fibrinous filaments and adhesions between the parietal and visceral pericardium. It finally may become thickened with abundant fibrous tissue with the possibility of forming loculations, as seen in our case.^{7,15} Other more specific but unusual findings such as typical rheumatoid nodules or vasculitis were absent in this patient.¹⁵

In conclusion, symptomatic RA pericarditis is a rare cardiac manifestation of RA, and a significant haemodynamic compromise is even rarer. The findings of the present case contribute to our

understanding of RA pericarditis. The case description illustrates that the condition might manifest with a giant compressive pericardial mass composed of fibrinous material, with particular involvement of the right ventricle. In such cases, exclusion of other conditions, such as neoplasms and infections, is necessary.

Lead author biography



Felipe Cañas is a current fellowship in Cardiology at ICESI University—Valle del Lili Foundation, Cali, Valle del Cauca, Colombia, Latin America. He was born in Manizales, Caldas, Colombia on 26 November 1987. He graduated from High School in Cali, Valle del Cauca, in June 2005, and got his Medical Degree from the Universidad del Valle in December

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Supplementary material

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

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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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