A 38-year-old man was admitted to the emergency department with a 12-hour onset of shortness of breath and position-related chest pain. In the last 3 months, he had had 2 similar but less intense episodes, each of which lasted for approximately 1 week. Both episodes were associated with subfebrility, flulike symptoms (dry cough, nasal obstruction), and increased fatigue and resolved with nonsteroidal antiinflammatory medication. The patient was diagnosed 2 years earlier with limited Wegener granulomatosis with upper and lower respiratory tract involvement (positive for antineutrophil cytoplasmic antibodies [anti-proteinase 3], a positive sinus ethmoidalis biopsy, and no renal involvement) and was undergoing immunosuppressive therapy (azathioprine and methylprednisolone). On admission, the patient was stable and afebrile and tolerated decubitus. The clinical examination was unremarkable, with no respiratory or cardiac pathological findings on auscultation. The chest radiograph showed a normal cardiac silhouette, clear lungs, and a small left pleural effusion. The ECG performed on admission showed sinus rhythm, normal PR interval, and diffuse flattened T waves. Initial blood tests showed mild anemia (hemoglobin 12.2 mg/dL) and marked inflammation, with an elevated erythrocyte sedimentation rate (86 mm/h) and C-reactive protein levels (306 mg/L). Troponin I on admission was within normal limits (0.02 μg/L). Given the clinical picture (nasal obstruction) and the laboratory results (high C-reactive protein and an increase in antiproteinase 3 compared with 2 months earlier, from 24 to 128 U/mL), there was a strong suspicion of relapsing disease. Transthoracic echocardiography showed a thickened pericardium with a circumferential, homogeneous pericardial effusion (up to 14 mm along the left ventricular [LV] lateral wall; Figure 1; online-only Data Supplement online-only Data Supplement Movie I) without a filling impairment (mitral valve: E inspiration 75 cm/s, E expiration 97 cm/s, 23% variation, E wave deceleration time 166 ms; Figure 2) and nondilated left and right chambers with normal biventricular systolic function. No valve abnormalities, pulmonary hypertension, or arguments for endocarditis were found. Thoracic computed tomography with contrast showed mild cardiomegaly, discrete left posterobasal pleural effusion, and thickened pericardium without calcifications (Figure 3). No lung infiltrates or adenopathies were present. For further characterization of the pericardial involvement and to exclude the presence of myocardial granulomatous infiltrates, cardiac magnetic resonance imaging was performed. Functional analysis by steady state free-precession cine sequences showed mildly dilated left and right ventricles (end-diastolic LV volume 124 mL/m² [normal values 79±8.7 mL/m²], right ventricular volume 109 mL/m² [normal values 88±11.7 mL/m²]), with mildly impaired LV systolic function (LV ejection fraction 50%) and normal right ventricular systolic function (online-only Data Supplement Movie II).

Figure 1. Transthoracic echocardiography. Midventricular short-axis (A) and parasternal long-axis (B) views show thickened pericardium (white arrowheads) with pericardial effusion (black arrowheads).

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Real-time cine imaging during free breathing showed minor inspiratory septal flattening but no septal inversion or shift (online-only Data Supplement Movie III), which argues against pericardial constriction. Morphological analysis by T1-weighted sequences showed normal appearance of the myocardium and thickened pericardial layers (up to 6 mm) with a hyperintense circumferential pericardial effusion (up to 7 mm along the LV lateral wall) that persisted after fat suppression, which suggests the presence of blood or a protein-rich exudate (Figures 4A and 4B). Edema imaging by T2-weighted short-τ inversion recovery (STIR) imaging showed intense circumferential edema of both pericardial layers and limited subepicardial edema in the inferolateral LV wall (Figure 4E). Late post–gadolinium administration imaging showed strong enhancement of both pericardial layers and subtle subepicardial enhancement of the inferolateral LV wall (Figures 4C and 4F). No valve abnormalities were seen.

Cardiac involvement in Wegener granulomatosis occurs in 6% to 44% of cases and, as is the case in other organs, is secondary to necrotizing vasculitis with granulomatous infiltrates. Pericarditis and coronary vasculitis are the most frequent findings (50% of cases), but myocarditis, endocarditis, and conduction system granulomata are also described. Currently, serological testing (antineutrophil cytoplasmic antibodies directed toward proteinase 3 with or without myeloperoxidase) and multiple available imaging modalities are increasingly used as first-line noninvasive diagnostic tools to assess target-organ involvement in Wegener granulomatosis, although biopsy with pathological testing represents the gold standard. We present a case of limited Wegener granulomatosis in which the diagnosis of cardiac involvement was made with several complementary imaging modalities. Magnetic resonance imaging was the most comprehensive, allowing us to appropriately describe the morphological correlate of the pericardial abnormalities, depict the presence of a mild myocarditis in the inferolateral LV wall, and accurately quantify ventricular volumes and function. This opens perspectives to adequately follow up patients with Wegener granulomatosis in whom the diagnosis of cardiac involvement is made, particularly given that evolution toward constrictive pericarditis is not infrequent.

Disclosures

None.

References

Figure 3. Thoracic computed tomography with intravenous contrast administration. Transverse (A) and coronal (B) images show thickened pericardium (arrowheads).

Figure 4. Cardiac magnetic resonance imaging. T1-weighted transverse views without (A) and with (B) fat suppression show diffusely thickened pericardium (arrowheads) and hyperintense appearance of the pericardial effusion along the left ventricular lateral wall. Late gadolinium enhancement horizontal long-axis (C) and midventricular short-axis (F) views show intense enhancement of both pericardial layers (arrowheads) and limited subepicardial enhancement of the left ventricular inferolateral wall (arrow). Midventricular short-axis cine image at end diastole (D) shows thickened pericardium (arrowheads). Midventricular short-axis T2-weighted short-TE inversion recovery image (E) shows edema of both pericardial layers (arrowheads) and limited edema of the left ventricular inferolateral wall (arrow).
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