

Cardiac Involvement in Erdheim-Chester Disease Magnetic Resonance and Computed Tomographic Scan Imaging in a Monocentric Series of 37 Patients

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Erdheim-Chester disease (ECD) is a rare non-Langerhans form of histiocytosis, characterized by the xanthomatous infiltration of tissues with foamy CD68⁺/CD1a⁻ histiocytes. By January 2009, >320 cases had been published in the medical literature. Bone pain is the most frequent symptom. Approximately half of patients have extraskeletal manifestations including exophthalmos, xanthelasma, interstitial lung disease, retroperitoneal “fibrosis” with perirenal or ureteral obstruction, renal failure, diabetes insipidus, and central nervous system and cardiovascular involvement. Interferon- α is the recommended first-line therapy but is decided after a case-by-case analysis.¹

Cardiovascular manifestations of ECD are underdiagnosed, as we have shown in our analysis in 2004 of the 178 cases known at that time.² Among these, we analyzed 72 patients with cardiovascular involvement and found 54 (75%) with heart involvement: pericardial infiltration in 32 patients (44%) (leading to tamponade in 5 cases), myocardial infiltration in 22 cases (31%), a right atrial tumor in 6 patients, and a symptomatic valvular heart disease in 6 patients (3 aortic and 3 mitral regurgitations); 19 patients (26%) had heart failure, leading to death in 8 cases; myocardial infarction (MI) was reported in 6 cases, leading to death in 2. Forty of the 72 patients (56%) had a periaortic fibrosis, and 20 of these had a “coated aorta” aspect. Among the 58 patients (81%) with available follow-up, 35 (60%) died. Death was due to the cardiovascular involvement in 31% of the cases, confirming the severe prognosis of ECD with cardiovascular complications. The poor prognosis of ECD with cardiovascular involvement led us to systematically search for it. Frequency and pattern of cardiac involvement in ECD, detected by magnetic resonance imaging (MRI), gated computed tomographic (CT) scan of the heart, or both, which were thus far unknown, are presented in this report.

Thirty-seven patients with ECD who were referred to the internal medicine department of our hospital between 1996

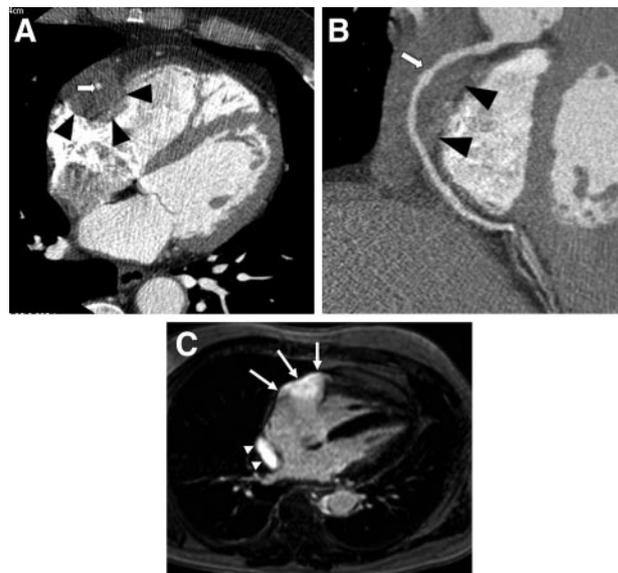


Figure 1. ECG-gated, contrast-enhanced, multislice CT. A, Axial slice shows a mass (black arrowheads) surrounding the right coronary artery (white arrow). B, Curved multiplanar reformat reveals the extension along segment II of the right coronary artery. C, Late-enhancement cardiac MRI shows strong enhancement of a mass located in the right atrioventricular sulcus (white arrows) and at the posterior wall of the right atrium (white arrowheads).

and September 2008 had a heart MRI, CT scan, or both. There were 25 male and 12 female patients, with a mean age at diagnosis of 52.3 years (range, 16 to 77 years). The analysis of the 37 chest x-rays showed a mild cardiomegaly in 2 patients and signs of right atrial enlargement in 3 patients. The aortic arch was enlarged in 1 patient. ECG data were available for 32 of the 37 patients. The ECG was normal in 12; remarkable findings were a short PR in 4 patients with in 1 case a PR at 7/100 with a sinoauricular block requiring a pacemaker, a sinus bradycardia in 3 other patients (in the absence of β -blockers), 2 more patients with ECG signs of

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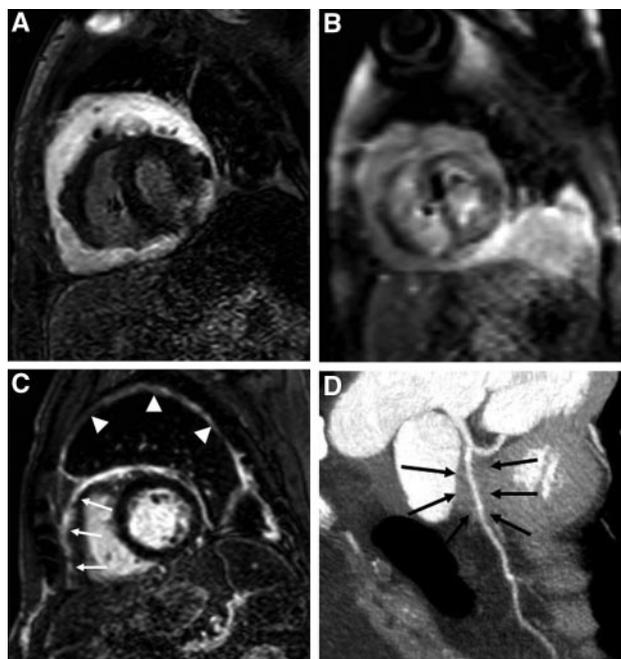


Figure 2. A, Late-enhancement cardiac MRI shows strong enhancement of both thickened pericardial layers. B, Still of short-axis real-time movie MRI during operator-guided breathing shows septal flattening during onset of inspiration, demonstrating pericardial constriction. C, Late-enhancement cardiac MRI shows strong enhancement of both thickened pericardial (white arrows) and pleural layers (white arrowheads). D, Curved planar reformation of a 64-detector computed coronary angiography shows large soft-tissue mass surrounding the proximal part of the left anterior descending artery (black arrows).

left ventricular hypertrophy, 3 patients with Q-wave abnormalities compatible with a MI (but no history of MI), and 2 more patients with both Q waves and a clinical history of MI: One patient had ST-T abnormalities (from V₁ to V₄), and 1 had a slight ST elevation (<1 mm) in D₁, D₂, D₃, VF, V₅, and V₆ with no past or present clinical history of pericarditis. Four patients had a pacemaker.

Twenty-five (68%) of the 37 patients had MRI of the heart, and 31 (84%) had an injected chest CT scan, 25 of which had a heart-gated examination (68%). Sixteen patients (43%) had both heart MRI and gated CT scan. Five patients (14%) had contraindications for MRI: Four had pacemakers or defibrillators, and 1 had a ventriculoperitoneal shunt.

In the present study, 26 patients (70%) had abnormal heart imaging. Eighteen patients (49%) exhibited abnormal infiltration of the right heart, 11 of whom (30%) had a “pseudo-tumoral” infiltration of the right atrium (Figure 1A and 1B), whereas 7 (19%) had an infiltration of the auriculoventricular sulcus (Figure 1C). Other remarkable findings were pericardial effusion in 9 patients (24%), pericardial thickening in 5 patients (14%) (Figure 2A) (1 with pericardial constriction [Figure 2B and Movie in the online-only Data Supplement] and 1 with gadolinium enhancement on MRI [Figure 2C]), periarterial infiltration of the left coronary artery in 10 patients (27%) (Figure 2D), and periarterial infiltration of the right artery (in the absence of pseudo-tumoral infiltration of the right atrium or of the auriculoventricular sulcus) in 2 patients (5%).

Our series, which is to date the largest for ECD, illustrates the benefit of a systematic screening for cardiac infiltration. One of the striking findings of ECD is the high frequency of right atrial and auriculoventricular sulcus involvement. Infiltration of the right heart has been classically described in angiosarcoma³ and lymphoma.⁴ The pericardial thickening, which may lead to tamponade, the periarterial coronary infiltration (with 7 MI, 2 of which died), and the “pseudo-atrial” mass are notably very well seen on heart imaging. A systematic cardiac evaluation by MRI, CT scan, or both should be performed in ECD patients because these manifestations are not always clinically evident.

Disclosures

None.

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