Magnetic Resonance Imaging Guiding Pacemaker Implantation for Severe Sinus Node Dysfunction Due to Cardiac Involvement in Erdheim-Chester Disease

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Magnetic Resonance Imaging Guiding Pacemaker Implantation for Severe Sinus Node Dysfunction Due to Cardiac Involvement in Erdheim-Chester Disease

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A 64-year-old woman was referred to the arrhythmia outpatient clinic after she had experienced syncope without preceding symptoms and frequent paroxysmal, near syncopal episodes over the last 8 months. The ECG revealed alternation of sinus bradycardia and frequent ectopic atrial beats, normal PR interval and right bundle branch block with consecutive repolarization abnormalities. Holter ECG showed frequent periods of asystole up to 4.3 seconds. Laboratory findings were normal. Pacemaker therapy for symptomatic sinus node dysfunction was clearly indicated.

The patient's past history revealed the diagnosis of Erdheim-Chester disease with osseous, cutaneous, mesenteric, and right atrial involvement 22 years ago. This disease belongs to a rare group of non-Langerhans cell histiocytosis of unknown origin. Tissue is infiltrated by foamy histiocytes.[1] The patient showed nearly pathognomonic radiographic changes in both tibiae and femora with bilateral symmetrical osteosclerosis of metaphyseal and diaphyseal regions with sparing of the epiphyses (Figure 1A).[2]

Over the last 5 years the patient has been stable through daily oral administration of 5 mg prednisolone.

Cardiac magnetic resonance imaging was performed to guide placement of the pacemaker electrodes and visualize the overall and local extent of tumor due to the known right atrial involvement by Erdheim-Chester disease. Echocardiographic findings are shown in Figure 1B.

Nearly complete and extensive involvement of the right atrial myocardium with thickening, septal bulge, and typical perivascular tumor spread[3] along the right coronary artery could be demonstrated (Figures 2A and 2B). Only a small area anterolateral of the right atrial wall was found to be less thickened, and showed no delayed hyperenhancement (Figures 2C and 2D). This area was chosen for atrial lead placement. Excellent pacing and sensing properties could be found.

Disclosures

None.

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The online-only Data Supplement, which contains Movies I and II, can be found at http://circ.ahajournals.org/cgi/content/full/115/16/e412/DC1.

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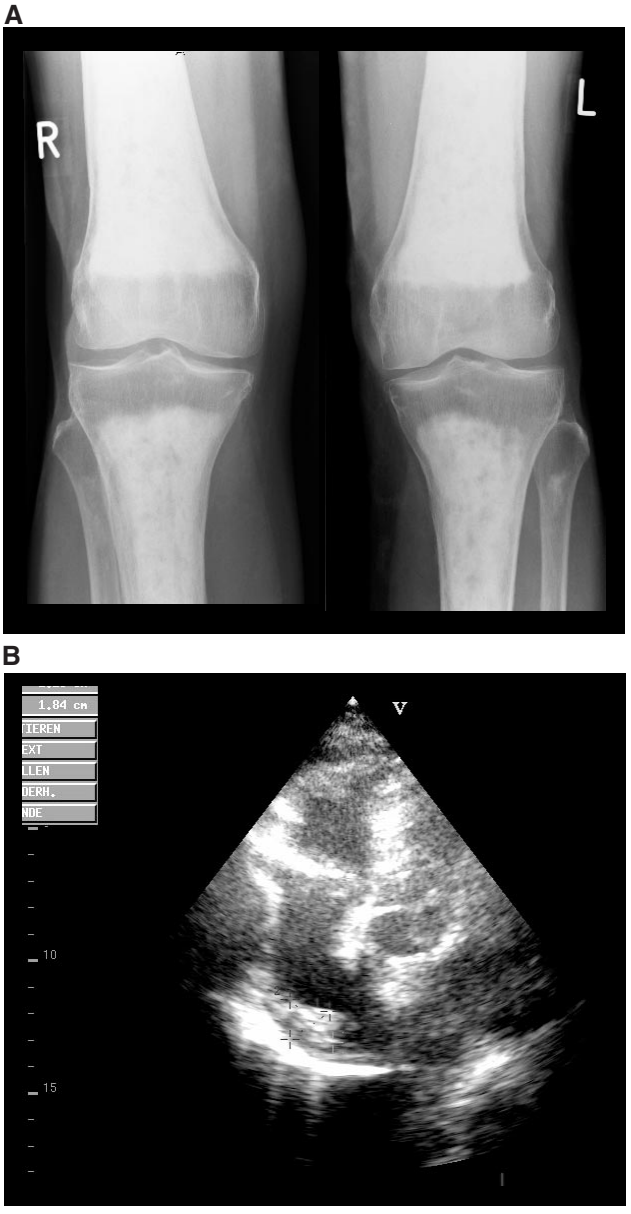

Figure 1. A, X-rays of both knees showing bilateral symmetric osteosclerosis of metaphysis with sparing of the epiphysis. B, echocardiography from an atypical apical view showing thickening of the right atrial wall (see measurements) and sparing of the lateral wall.

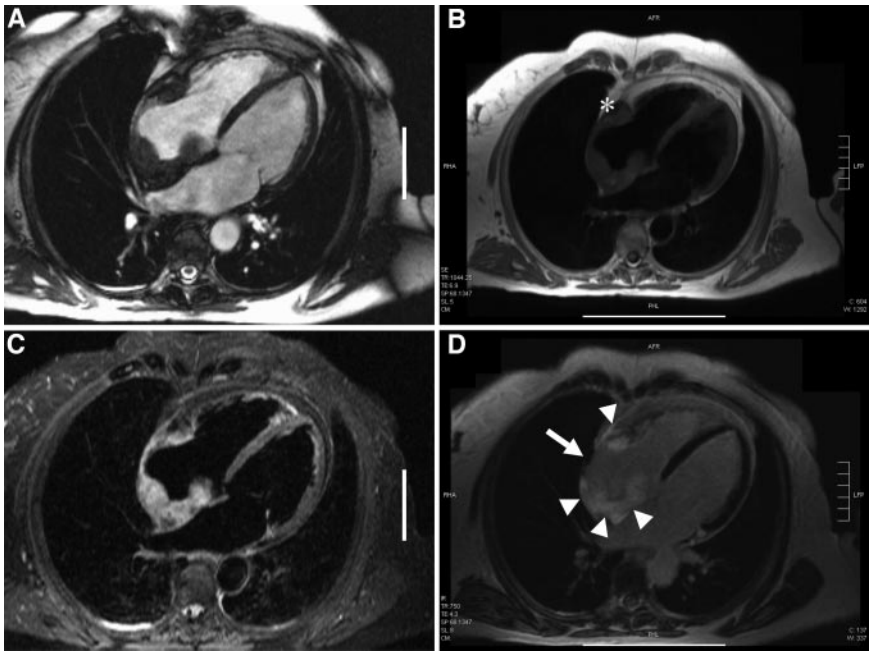

Figure 2. A, Four-chamber view of the heart using Cine-SSFP sequence (echo time 1.57 ms, repetition time 47.1 ms, 5 mm slice thickness). B, T1 weighted imaging (echo time 6.9 ms, repetition time 1044 ms, 5 mm slice thickness) in same orientation. The thickening of right atrial wall and septal bulge can clearly be seen. Typical perivascular tumor spread (asterisk) along the right coronary artery. C, The bright signal in T1 weighted images shows a signal decay after fat suppression (echo time 62 ms, repetition time 2110 ms) suggesting partially fat-containing parts of the tumor. D, T1 weighted imaging 10 minutes after contrast material application (0.2 mmol per kg body weight Gadodiamide) with suppression of normal myocardium (black) shows extensive delayed hyperenhancement indicative of granulomatosis/fibrosis in the thickened right atrial wall (echo time 4.3 ms, repetition time 750 ms, inversion time 230 ms: arrowheads). A small anterolateral area of the right atrial wall is less thickened and shows no delayed hyperenhancement (arrow). No ventricular delayed hyperenhancement was seen; so there was no evidence of myocardial infarction.