Case Reports

Multidetector Computed Tomographic Imaging of Erdheim-Chester Disease

Zeyneb Yuceler, MD
Mecit Kantarci, MD, PhD
Nevzat Karabulut, MD
Hayri Ogul, MD
Ummugulsum Bayraktutan, MD
Canan Akman, MD

Erdheim-Chester disease is a rarely reported disease that can affect nearly every organ and chiefly infiltrates the connective, perivascular, and adipose tissue. The disease is a form of non-Langerhans-cell histiocytosis characterized by the proliferation of foamy histiocytes; its cardiovascular complications carry a severe prognosis. We present the case of a 29-year-old woman who was admitted for analysis of her angina. Our evaluation with use of cardiac multidetector computed tomographic angiography revealed large mediastinal soft tissue that compressed the patient's left anterior descending coronary artery. To our knowledge, this is the first report of the use of low-dose, dual-source, 256-slice multidetector computed tomography to characterize Erdheim-Chester disease that exclusively caused angina and stenosis of a coronary artery in a young adult. ***(Tex Heart Inst J 2014;41(3):338-40)***

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From: *Department of Radiology (Drs. Bayraktutan, Kantarci, Ogul, and Yuceler), School of Medicine, Ataturk University, 25000 Erzurum; Department of Radiology (Dr. Karabulut), School of Medicine, Pamukkale University, 20000 Denizli; and Department of Radiology (Dr. Akman), Cerrahpasa Medical Faculty, Istanbul University, 34000 Istanbul; Turkey*

Address for reprints: *Mecit Kantarci, MD, 200 Evler Mah. 14. Sok No:5, Dadaskent, 25090 Erzurum, Turkey*

E-mail: *akkanrad@hotmail.com*

Erdheim-Chester disease (ECD) is a rarely reported form of multisystemic, non-Langerhans-cell histiocytosis. The clinical manifestations range from no symptoms to life-threatening conditions. Involvement of the bones, lungs, retro-orbital space, perirenal space, nervous system, cardiovascular system, and skin has particularly been observed. The cardiovascular involvement of ECD is probably more frequent than was originally thought; technological advances in multidetector computed tomography (MDCT) have enabled better views of patients' coronary arteries at low radiation doses. We report the case of a young woman in whom we used 256-slice, dual-source MDCT with high pitch value to evaluate the coronary arteries and reveal the effects of ECD.

Case Report

In April 2013, a previously healthy 29-year-old woman was referred to our hospital with a 3-month history of progressive angina. She had no history of other medical illnesses or familial diseases. Results of physical examination, laboratory tests, electrocardiography (ECG), and echocardiography were normal. To analyze her coronary arteries, we used a 256-slice SOMATOM® Definition Flash MDCT system (Siemens Medical Solutions; Forchheim, Germany) with a high pitch value of 3.2. The patient's effective radiation dose was 1.1 mSv for the cardiac imaging. The MDCT angiogram revealed large mediastinal soft tissue with muscle attenuation surrounding the ascending aorta, extending into the proximal segment of the left main coronary artery, and causing stenosis by compressing the proximal segment of the left anterior descending coronary artery (Figs. 1 and 2). This periaortic effect resembles "coated aorta," associated with ECD. A specimen from a computed tomographic-guided biopsy of the mediastinal mass displayed fibrosis and many foamy histiocytes ($CD68^+$ and $CD1a^-$). The imaging and histologic findings supported the diagnosis of ECD. Skeletal radiographs showed no bone involvement, and thoracoabdominal computed tomograms and magnetic resonance images of the brain and orbit revealed no effect on those organs. The patient was referred for hemato-oncologic treatment. She was subsequently lost to follow-up.

Discussion

To our knowledge, this is the first report of the use of low-dose, dual-source, 256-slice multidetector computed tomography to characterize Erdheim-Chester disease

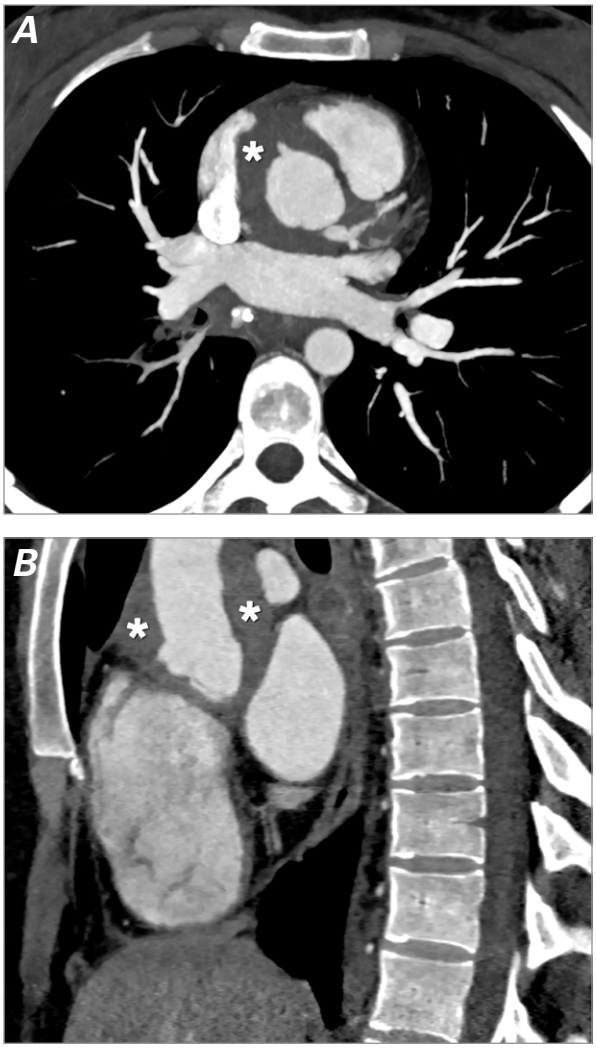

Fig. 1 *Multidetector computed tomographic images show a soft-tissue mass (asterisks) surrounding the ascending aorta* ***A****) in axial maximum-intensity projection and* ***B****) in the sagittal plane.*

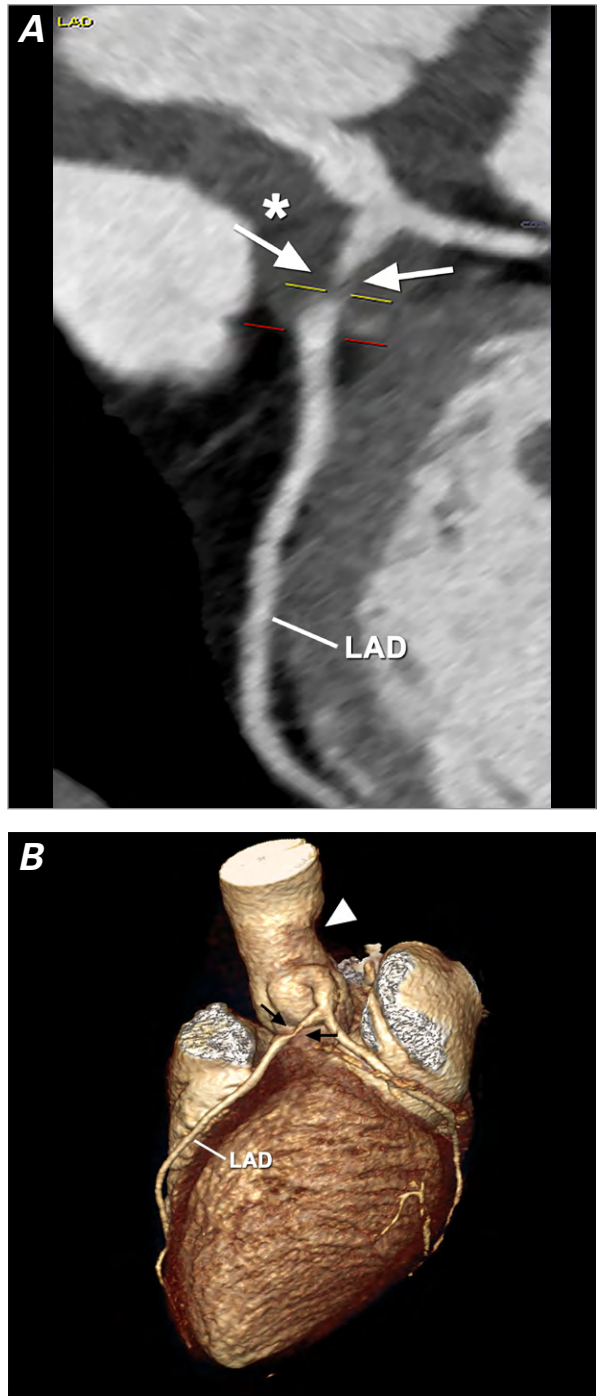

Fig. 2 *Multidetector computed tomographic images.* ***A****) Reformatted curved planar image shows a soft-tissue mass (asterisk) causing stenosis (arrows) of the left anterior descending coronary artery (LAD).* ***B****) Three-dimensional reconstruction shows compression of the aorta (arrowhead) and stenosis of the LAD (arrows).*

that exclusively caused angina and stenosis of a coronary artery in a young adult.

Erdheim-Chester disease is an uncommon form of multisystemic non-Langerhans-cell histiocytosis. In ECD, lipid-containing foamy histiocytes proliferate and infiltrate connective, perivascular, and adipose tissue. This disease was first described in 1930 as a form of lipogranulomatosis with bone and pericardial involvement.[1] However, there is no demonstrated association with lipid metabolism abnormalities.[2] Erdheim-Chester disease affects females and males of any age, with a minor male predilection after the 4th decade of life.[3]

This disease can affect nearly every organ and particularly infiltrates the long bones, resulting in xanthogranulomatous tissue that leads to bilateral osteosclerosis, which is a pathognomonic radiologic sign of ECD. In addition, ECD can lead to interstitial lung disease and dyspnea by affecting the interlobular septa and pleura, diabetes insipidus by infiltrating the pituitary gland, hydronephrosis and renal failure by invading the retroperitoneal connective and adipose tissue, exophthalmos by occupying the retro-orbital space, demyelinating responses in the central nervous system, and xanthelasma

in the skin. Infiltration of the cardiovascular system can manifest itself as pericardial effusion, pericardial constriction, tamponade, congestive heart failure, right atrial pseudotumoral infiltration, valvular disorders, myocardial infarction due to coronary artery involvement, periaortic fibrosis, and renovascular hypertension due to renal artery stenosis.[4] Although the cardiovascular complications have a severe prognosis in ECD patients,[5] the symptoms can be subclinical and be overlooked unless a systematic cardiac evaluation is conducted.[5,6]

In ECD, the mediastinal infiltration often starts around the aorta and aortic branches. As a result, the most frequent cardiovascular evidence is periaortic circumferential tissue infiltration, known as a coated aorta.[7-9] The aortic effects and fibrosis can extend from the ascending aorta down to the iliac bifurcation or be limited to the thoracic or abdominal aorta.[5] Unlike Takayasu arteritis and retroperitoneal fibrosis, ECD infiltrates the periadventitial or perivascular area circumferentially, regularly forming a tissue with the same attenuation as muscle. (Nevertheless, the intima might appear to be irregular in computed tomographic images.[8]) In contrast, in Takayasu arteritis, the infiltration is parietal and extends throughout the layers of the aortic wall. The posterior part of the aorta is also maintained, and the inferior vena cava is generally affected by retroperitoneal fibrosis.[3,10] Periaortic and coronary artery effects alone can be observed in ECD, as in our patient. Accordingly, ECD should be kept in mind as a rare cause of angina, particularly in patients with external compression of the coronary arteries.

Treatment for ECD varies in accordance with clinician preference: systemic steroids, various cytotoxic agents, radiation therapy, and hematopoietic stem cell transplantation have been attempted.[11,12] Video-assisted thoracoscopic surgery should be used for diagnosing cardiac effects and managing recurrent pericardial effusion in ECD.[13]

Fewer than 45 cases of ECD have been reported in association with coronary artery involvement.[5,8,9,14-16] The cardiovascular manifestations of ECD are being detected more frequently because of improved imaging techniques, indicating that the number of such cases has been underestimated. Currently, the best images and low radiation doses are produced by 256-slice, dual-source MDCT, prospectively ECG-triggered acquisition, and a high pitch value of 3.2 or 3.4.[17] We found that low-dose, 256-slice, dual-source MDCT readily enabled evaluation of the coronary arteries and mediastinum in our young female patient with ECD.

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