

Imaging Assessment of Periaortic Inflammation in Erdheim-Chester Disease

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Abstract

Reaching etiologic diagnoses for retroperitoneal fibrosis may be challenging. We report the case of a 75-year old male with history of ruptured abdominal aortic aneurysm and subsequent retroperitoneal fibrosis who developed four years later a soft tissue infiltration surrounding the ascending thoracic aorta. Thanks to his medical records and multimodality imaging assessment, the patient escaped an open-chest biopsy through histological reassessment of the abdominal periaortic samples that allowed the definitive diagnosis of Erdheim-Chester disease, a rare non-Langerhans histiocytosis.

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Key Words

Aortitis · Histiocytosis · Erdheim-Chester · Magnetic Resonance Imaging · Positron Emission Tomography

Case Report

A 75-year-old male presented with three self-subsiding episodes of malaise and nausea. Medical history included surgery for ruptured inflammatory abdominal aortic aneurysm, four years earlier; pathology of the surgical specimen showed idiopathic perianeurysmal fibrosis. Postoperative follow-up was marked by periaortic graft infiltration involving both ureters, necessitating bilateral ureteral stenting and steroid therapy for eight months. The patient had remained asymptomatic for three years until the onset of malaise. Clinical examination and

ECG were unremarkable. The serum C-reactive protein was at 7 mg/L (normal 0–6), and blood cell count showed neutrophilia (85%) without hyperleucocytosis and a slight microcytic anemia (hemoglobin: 12.4 g/100 mL). On echocardiography, a 10 mm pericardial effusion (PE) was noticed and subsequent computed tomography (CT) displayed a layer of abnormal soft tissue surrounding the thoracic aorta (Figure 1A, arrows), which was not present on a prior examination (not shown). Whole-body transverse ¹⁸F-fluoro-deoxyglucose positron emission tomography (FDG-PET) (Figure 1B) and high b-value (800 s/mm²) diffusion-weighted MRI (DW-MRI) (Figure 1C) were performed and, respectively, color map-fused with CT (Figure 1D and 1E), showing in a roughly similar distribution, an increased FDG uptake and decreased water diffusion (arrows). No other pathological area was identified. Both molecular imaging techniques helped with differentiating multiorgan malignancy from perivascular inflammatory diseases, allowing hypothesis that the current disease and the previous perianeurysmal fibrosis are actually two expressions of the same disease. We, therefore, discussed either Erdheim–Chester disease, or large vessel vasculitis such as Takayasu and giant cell arteritis. Erdheim–Chester disease is an uncommon non-Langerhans cell histiocytosis characterized by a perivascular, adipose, and connective tissue tropism [1–3] that may be responsible for widespread vascular involvement, including “chronic periaortitis,” a ge-



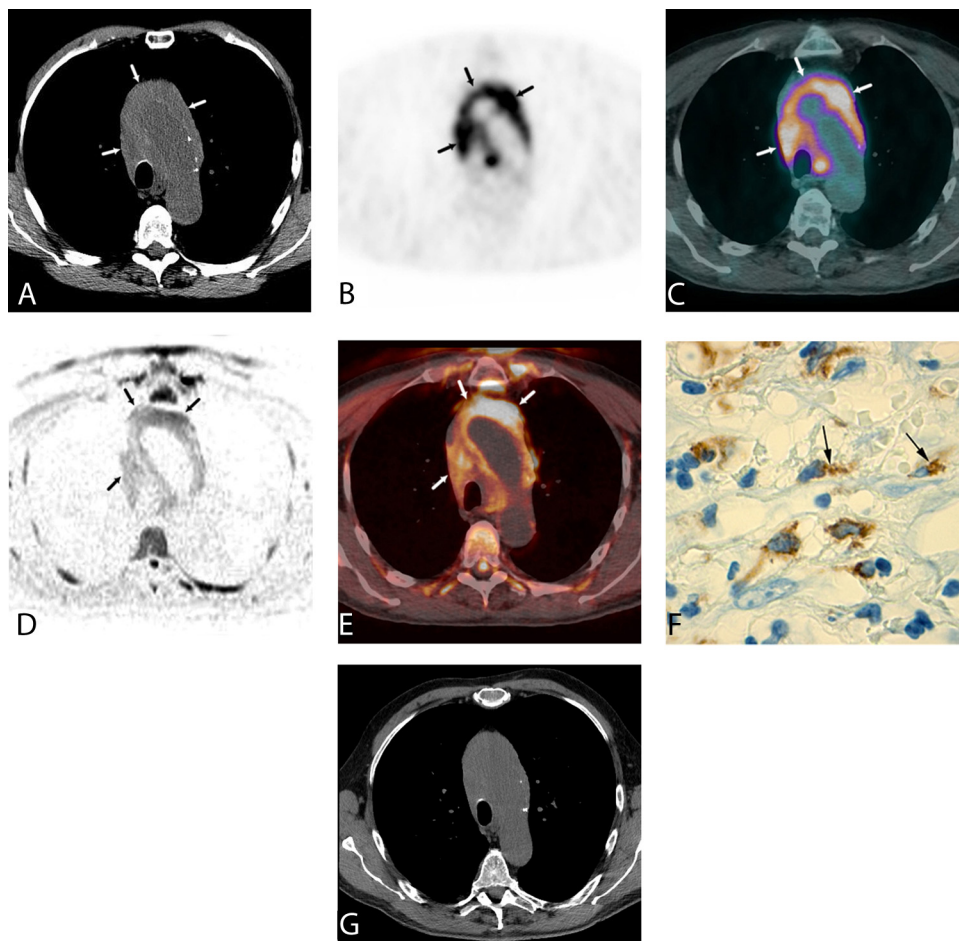


Figure 1. A 75-year-old male with malaise and history of inflammatory abdominal aortic aneurysm rupture four years earlier. (A) Unenhanced transverse computed tomography (CT) of the chest demonstrates a perivascular soft tissue mass, encasing the aortic arch (**arrows**). (B) ¹⁸F-Fluorodeoxyglucose positron emission tomography (FDG PET) and (C) color intensity map fusion with CT showed a remarkable uptake of the tissue. (D) Transverse diffusion-weighted magnetic resonance with a diffusion-factor value of 800 s/mm² and intensity color maps fusion to CT (E) images showed restricted diffusion (**arrows**) in a roughly identical distribution to FDG uptake. Histological reassessment of the samples obtained during abdominal aortic surgery (F) demonstrates inflammatory foci with predominance of foamy cells (histiocytes) infiltrates with cytoplasmic brown deposition at immunoperoxidase stain with CD68 (**arrows**), consistent with the diagnosis of Erdheim–Chester disease for which other immunohistological hallmarks were a negative staining for both CD1a and S-100 protein (not shown). (G) Unenhanced transverse CT showing mild decrease of the perivascular soft tissue after treatment.

neric term for perianeurysmal retroperitoneal fibrosis, inflammatory abdominal aortic aneurysm, and idiopathic retroperitoneal fibrosis. This tropism noticed in our patient history was a clue to the diagnosis, even though the initial pathological evaluation may have failed for several reasons, including undersampling. We proceeded to a histopathologic reassessment of the perianeurysmal samples obtained 4 years earlier. It revealed inflammatory foci with a predominance of foamy cells (histiocytes) infiltrates with cytoplasmic

brown deposition at immunoperoxidase stain with CD68 (Figure 1F, arrows) that eventually allowed the definitive diagnosis of Erdheim–Chester disease. Other immunohistological hallmarks of the disease were a negative staining for both CD1a and S-100 protein (not shown). The patient was treated by further steroid administration of methylprednisolone. Follow-up chest CT showed a marked decrease of the periaortic infiltration (Figure 1G), while the patient remained asymptomatic.

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