A 47-year-old man with a history of gynecomastia due to prolactin-releasing pituitary microadenoma was admitted to the hospital because of persistent chest pain and fatigue. Chest x-ray showed pronounced enlargement of the cardiac profile (Figure 1), and echocardiography demonstrated a marked pericardial effusion (Figure 2). Pericardiocentesis allowed the drainage of 1800 mL of pericardial fluid whose biochemical analysis was consistent with an exudate (total protein and lactate dehydrogenase fluid-to-serum ratios of 0.9 and 2.6, respectively); cytological examination showed the presence of mesothelial cells, histiocytes, and lymphocytes.

Whole-body computed tomography disclosed the presence of a homogeneous mass of soft-tissue density coating the thoracic (Figure 3) and abdominal aorta (Figure 4) and also surrounding both kidneys (Figure 4). Computed tomography-guided biopsy of the perirenal masses was performed, and histology showed a diffusely fibrotic tissue with a population of infiltrating histiocytes, which stained positive for CD68 (Figure 5) and negative for S100 and CD1a (data not shown). These findings were consistent with Erdheim-Chester disease (ECD), a diagnosis that was also supported by the presence of osteosclerotic lesions involving the tibia.

ECD is a rare form of non-Langerhans cell histiocytosis whose hallmark is tissue infiltration by CD68+CD1a-foamy non-Langerhans histiocytes and mononuclear cells, along with pronounced fibrosis. The mechanisms leading to accumulation of histiocytes in ECD lesions seem to be chemokine-mediated, although clonal histiocyte proliferation has also been demonstrated in some cases. ECD has a broad spectrum of clinical manifestations and may range from a localized pauci-symptomatic form to a disseminated and life-threatening disease; it typically affects the long bones, the hypothalamic-pituitary axis, the skin, the lungs, and the retroperitoneum. Cardiovascular involvement is considered to be an overlooked feature of ECD, although diffuse periaortic fibrosis (also known as “coated aorta”), pericardial infiltration with pericardial effusion, myocardial infiltration, atrial pseudotumour, and aortic and mitral valve disease are not uncommon clinical findings. A careful evaluation of cardiovascular involvement in ECD patients is warranted because it accounts for a considerable proportion of ECD-related deaths.

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Disclosures

None.

References

Figure 2. Transthoracic echocardiographic image, obtained on admission to hospital, showing large circumferential pericardial effusion (apical 4-chamber view). PE indicates pericardial effusion; LV, left ventricle; RV, right ventricle; LA, left atrium; and RA, right atrium.

Figure 3. Contrast-enhanced computed tomography scan of the chest showing the presence of muscle-isodense tissue surrounding the thoracic aorta, particularly the ascending portion (arrow).

Figure 4. Contrast-enhanced computed tomography scan of the abdomen showing muscle-isodense tissue encircling the abdominal aorta (arrow) and also surrounding both kidneys (arrowheads).

Figure 5. A, Low-power view of the needle biopsy specimen at the boundary between the renal cortical parenchyma (asterisk) and the perirenal retroperitoneal soft tissues. The latter are diffusely fibrotic and infiltrated by cells frequently organized in clusters (arrows). Hematoxylin-Eosin stain; original magnification ×4; scale bar is 400 μm. B, The infiltrating elements (arrows) are mononuclear, show either polygonal or elongated shape, and diffusely permeate the collagen fibers (reddish bands). The mild inflammatory background is mainly composed of small lymphocytes (arrowheads). Hematoxylin-Eosin stain; original magnification ×20; scale bar is 100 μm. C, Immunohistochemically, the mononuclear cells show strong cytoplasmic granular positivity for the CD68 antigen (brown color), thus indicating a histiocyte phenotype. Counterstained with Mayer hematoxylin, original magnification ×20; scale bar is 100 μm.
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