

# The Hairy Kidney of Erdheim-Chester Disease



Jack C. Scolaro, MS, and Alan N. Peiris, MD, PhD

A 63-year-old man with a medical history of osteoarthritis, gastroesophageal reflux disease, and myocardial infarction presented with upper abdominal pain of several months duration. Pain was worse with palpation. Upper and lower endoscopy revealed mild gastritis and the patient was given a trial of pantoprazole. On follow-up, a computed tomography (CT) of the abdomen and pelvis was ordered because of lack of improvement. The CT showed abnormal thickening around the kidneys, adrenals, and descending aorta. The renal involvement seen in this image (Figure) displays a unique “hairy kidney” appearance commonly associated with Erdheim-Chester disease (ECD),<sup>1</sup> an extremely rare condition with approximately 750 reported cases in literature.<sup>2</sup>

Erdheim-Chester disease is a non-Langerhans cell histiocytosis characterized by the infiltration of foamy histiocytes with fibrosis that can affect many organs, most commonly long bones.<sup>3</sup> Renal involvement commonly manifests in this disease as a histiocytic infiltration into the perirenal fat (arrows). Histopathologic findings from biopsy of the right retroperitoneal region included fragments of fibroadipose tissue displaying a lymphohistiocytic infiltrate. These histiocytes exhibited foamy cytoplasm with irregular nuclei. Touton-type giant cells were also noted. Immunohistochemical stain was positive for CD68, confirming the presence of histiocytes. In addition, the cells were negative for S100, ruling out Langerhans cell histiocytosis.<sup>4</sup> These findings enabled a clear diagnosis of ECD. There are no guidelines for treating ECD. Treatments include interferon- $\alpha$ , targeted chemotherapy with serine/threonine protein kinase B-raf (BRAF) inhibitors,<sup>5</sup> steroids, immunotherapy,



**FIGURE.** Abdominal computed tomography displays “hairy kidney” appearance associated with Erdheim-Chester disease.

From Texas Tech Health Sciences Center, School of Medicine, Lubbock, TX.

radiation, and surgery. The BRAF testing was inconclusive in this patient, and because of mild symptoms the patient was not started on chemotherapy.

**Potential Competing Interests:** The authors report no competing interests.

**Correspondence:** Address to Jack C. Scolaro, Texas Tech University Health Sciences Center, School of Medicine, 3601 4th St, Lubbock, TX 79430 ([jack.scolaro@ttuhsc.edu](mailto:jack.scolaro@ttuhsc.edu); [alan.peiris@ttuhsc.edu](mailto:alan.peiris@ttuhsc.edu)).

1. Campochiaro C, Tomelleri A, Cavalli G, Berti A, Dagna L. Erdheim-Chester disease. *Eur J Intern Med.* 2015;26(4):223-229.
2. Stempel JM, Bustamante Alvarez JG, Carpio AM, Mittal V, Dourado C. Erdheim-Chester disease, moving away from the orphan diseases: a case report. *Respir Med Case Rep.* 2016;20:55-58. eCollection 2017.
3. Veyssier-Belot C, Cacoub P, Caparros-Lefebvre D, et al. Erdheim-Chester disease: clinical and radiologic characteristics of 59 cases. *Medicine (Baltimore).* 1996;75(3):157-169.
4. Diamond EL, Dagna L, Hyman DM, et al. Consensus guidelines for the diagnosis and clinical management of Erdheim-Chester disease. *Blood.* 2014;124(4):483-492.
5. Haroche J, Charlotte F, Arnaud L, et al. High prevalence of BRAF V600E mutations in Erdheim-Chester disease but not in other non-Langerhans cell histiocytoses. *Blood.* 2012;120(13):2700-2703.