

Clinical Image

Hairy Kidney Sign a Must Know Diagnosis

Soukayna J*, Khadija ELA, Jamal EIF and Tarik S
Radiology Department, National Institute of Oncology,
Rabat, Morocco

***Corresponding author:** Jabour Soukayna, Radiology
Department, National Institute of Oncology, Rabat,
Morocco

Email: dr.jaboursoukayna@gmail.com

Received: July 14, 2025

Accepted: July 24, 2025

Published: July 25, 2025

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Keywords: Rare disease; CT imaging; Early diagnosis

Discussion

Erdheim-Chester disease (ECD) is a rare, acquired form of non-Langerhans cell histiocytosis affecting adults. It is characterized by xanthogranulomatous infiltration of lipid-laden histiocytes and presents with a wide range of systemic symptoms. The most common clinical features involve the skeleton—particularly the long bones—which often show increased uptake on technetium-99m bone scintigraphy [1], and the urinary tract, notably with retroperitoneal fibrotic changes. Diagnosis is typically confirmed through ultrasound-guided biopsy of the perirenal infiltrative tissue.

Urological manifestations are present in approximately 30% of ECD cases. These frequently appear as retroperitoneal fibrosis, which can progress to bilateral hydronephrosis and impaired renal function. In contrast to idiopathic retroperitoneal fibrosis (RPF), ECD-associated fibrosis tends to spare the ureters and does not involve the inferior vena cava. A distinguishing feature is the circumferential encasement of the aorta, whereas idiopathic RPF usually preserves the posterior aspect of the vessel. ECD should be considered in all patients presenting with corticosteroid-resistant retroperitoneal fibrosis. Special attention should be paid to the imaging appearance of the perirenal fascia and fat, which may exhibit the characteristic perirenal rind or "hairy kidney" sign (Figure 1).

References

1. William Jeffrey Triffo, Raymond B Dyer. The "hairy kidney" sign. Abdominal Radiology. 2017; 42: 979.
2. Kunin M. Bridging septa of the perinephric space: anatomic, pathologic, and diagnostic considerations. Radiology. 1986; 158: 361-365.

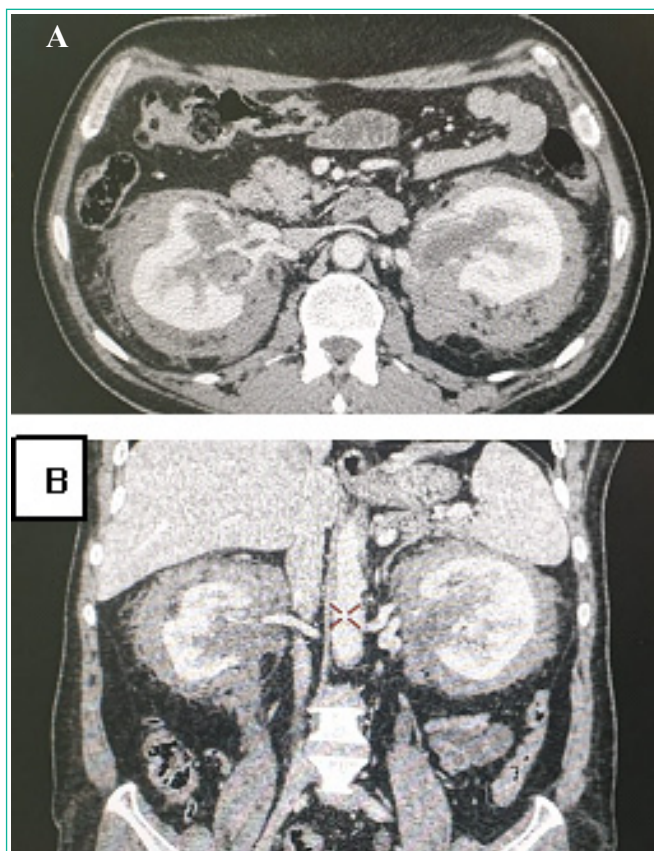


Figure 1: chromatogram of sample solution (80 µg/mL).