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Case Report

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Erdheim-Chester Disease: A Case Report with Radiographic Characteristics

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Introduction

Erdheim-Chester disease (ECD) is a rare non-Langerhans cell histiocytosis. The disease is highly variable in severity, ranging from incidental findings in asymptomatic patients to fatal multisystem disease [1]. Diagnosis is based on characteristic histological and radiological findings.

We report the case of an 85-year-old man who presented to our department with diffuse arthralgia and abdominal pain without fever. Laboratory tests showed no cytopenia, cytolysis or cholestasis and no biological inflammatory syndrome. Abdominal CT scan showed densification of peri-renal fat and the appearance of hairy kidneys. Standard x-rays of the long bones did not show any abnormalities. Bone scintigraphy showed no specific lesions. The diagnosis of ECD was suspected.

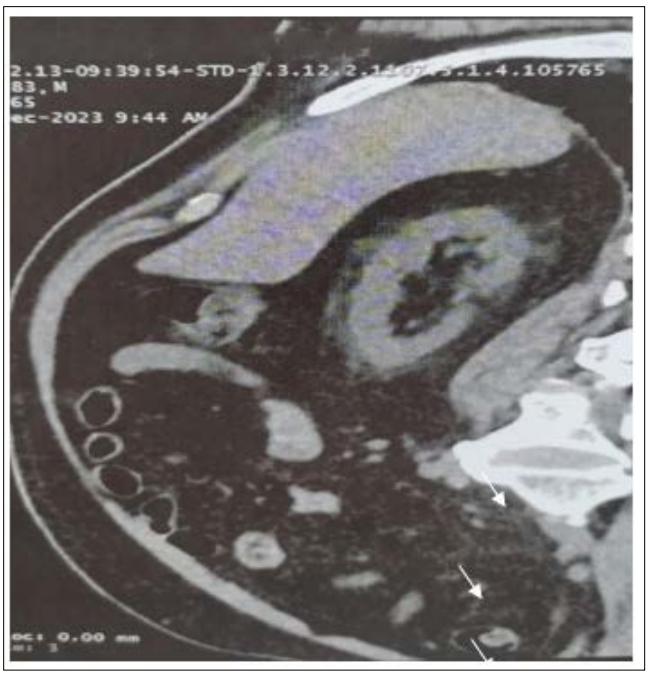

Figure 1: Abdominal CT scan (Longitudinal Section): symmetric infiltration of perirenal fat and perirenal fascia. Perirenal fat and fascia, giving a "hairy kidney" appearance.

Differential Disease

- Langerhans'cellhi Stiocytosis
- Retroperitoneal Fibrosis
- Rosai–Dorfman Disease

References

1. Benson JC, Vaubel R, Ebne BA, Mark IT, PerisCelda M, et al. (2023) Erdheim-Chester Disease. American Journal of Neuroradiology 44: 505-510.

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