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Perirenal fibrosis: make your diagnosis

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Case

A 72-year-old patient treated with BCG immunotherapy for relapse of urothelial cancer was admitted for diffuse oedema. His past history was characterized by recurrent pleuropericarditis treated by pericardotomy 16 years earlier and by retroperitoneal fibrosis (RPF) treated by steroids 15 years earlier. Serum creatinine rose progressively from 190 to 618 µmol/L and he was admitted in the nephrology department. Clinical examination showed diffuse oedema, dyspnoea and mild tibial tenderness.

Haemoglobin was 99 g/L with normal platelet and leukocyte counts. Calcaemia, phosphoraemia, magnesaemia, haptoglobin and lactate deshydrogenase levels were within the normal range. Serum was negative for antinuclear and antineutrophil cytoplasm antibodies.

Bilateral pyelic dilatation was diagnosed and treated by bilateral ureteral stenting.

Echocardiography, cardiac magnetic resonance imaging (MRI) and right cardiac catheterization confirmed restrictive cardiopathy related to constrictive pericarditis with severe thickening of the lateral wall of the pericardium (not shown). An abdominal CT scan was performed and is shown in Figure 1.

Diagnosis

The CT scan showed a symmetrical infiltration of the perirenal fat consistent with a 'hairy kidney' appearance prompting us to suspect Erdheim–Chester Disease (ECD). A strong uptake of the tracer by long bones was seen on the ^{99}Technecium bone scintigraphy (Figure 2A).

Histological analysis of the renal capsule showed fibrosis with infiltration composed of scattered histiocytes with clear cytoplasm (Figure 2B). These histiocytes expressed CD68 (Figure 2C) but not CD1a and PS100. The diagnosis of ECD was retained and treatment with pegylated-alpha (IFN-α) was started. Renal failure worsened and chronic dialysis was started. Past history of constrictive pericarditis and RPF were certainly the first manifestations of ECD. Unfortunately, pericardial histology was not available.

ECD is a very rare, non-Langerhans form of histiocytosis with a wide range of manifestations and a severe prognosis. ECD was first described as the 'lipoid granulomatose' by Jakob Erdheim's pupil, William Chester in 1930 [1]. A total of 445 cases had been reported until July 2011 [2]. RPF may be seen in ∼30% of ECD [2]. Tracer uptake by long bones on bone scintigraphy and the 'hairy kidney' appearance on CT-scan are almost always found and are the most evocative signs of ECD [2]. Although nearly all patients have osseous involvement, bone pain is present in only 50% of cases. Cardiovascular involvement is severe and is responsible for one third of the deaths. The central nervous system, especially the cerebellum, can also be affected [3]. Retro-orbital infiltration, endocrine and pulmonary involvement have also been reported [2]. Before the use of IFN-α, the mean survival time was 19.2 months (range: 0–120) [2]. More recent

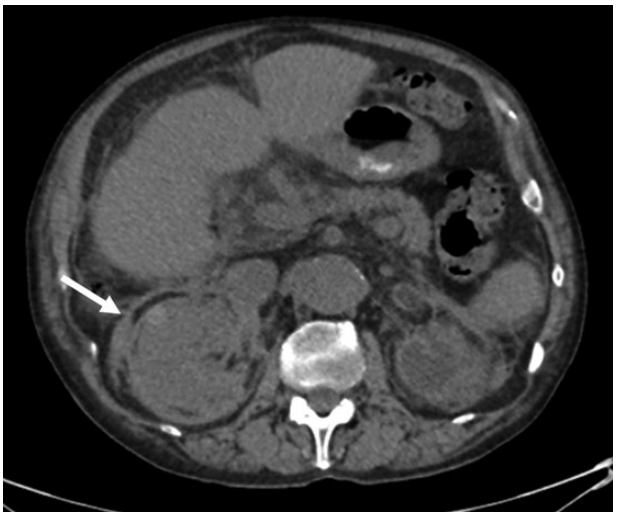

Fig. 1. Abdominal CT scan showing a symmetrical infiltration of the perirenal fat and of the perirenal fascia taking the appearance of 'hairy kidneys' (white arrow).

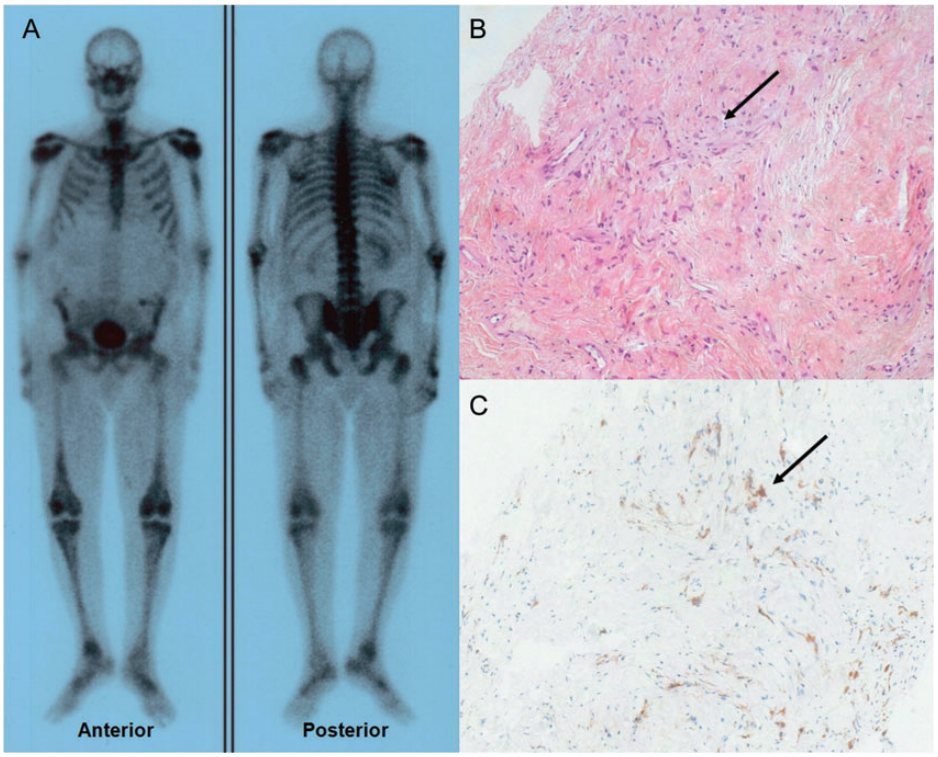

Fig. 2. Uptake of the tracer by long bones on the ^{99}Technecium bone scintigraphy. (**A**) Biopsy of the renal capsule, showing fibrosis with infiltration composed of scattered histiocytes with clear cytoplasm (**B**). Immunohistochemistry analysis showing the expression of CD68 by histiocytes (**C**). Magnification ×100.

analyses, after the use of IFN-α, showed a 5-year survival rate of 68% [2]. Recombinant interleukin-1 receptor antagonist (anakinra) was also used with success [4]. Recently, BRAF V600F mutation was detected in 54% of ECD [5], leading to the use of vemurafenib in three refractory patients that dramatically improved [6]. In the present case, BRAF V600F mutation was not detected.

ECD is a rare non-Langerhans histiocytosis that has to be considered by nephrologists in the case of RPF, especially when 'hairy kidneys' are seen on abdominal CT scan. Thus, specific treatments can apply to improve the course of disease for the patients.

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Conflict of interest statement. None declared.

(See related article by Sandal and Zand. Page kidney phenomenon secondary to an atypical presentation of Erdheim-Chester disease. *Clin Kidney J* 2013; 6: 547–548)

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