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Medical Imaging

Scalped Kidneys Fortuitous Discovery at Computed Tomography: About A Case in Bamako

Traore Ousmane[1,7*], N'diaye Mamadou[2], Diarra Ouncoumba[3], Thiam Awa[4], Coulibaly Modibo[4], Bagayoko Ousmane Lansenou[1], Guindo Ilias[5], Diallo. Mahamdou[6], Keita Adama Diaman[7]

[1]Medical Imaging Department of the ''Marie Curie'' Medical Clinic in Bamako-Mali
[2]Radiology Department of the Bamako-Mali Military Camp Health Center
[3]Radiology Department of the Reference Health Center of Commune III of Bamako-Mali
[4]Reference Center Service of Commune V of Bamako-Mali
[5]Department of Radiology of the University Hospital Center of Kati-Mali
[6]Gyneco-Obstetrics Department of the University Hospital Center "Gabriel TOURE" Bamako-Mali
[7]Radiology Department of the University Hospital Center of Point "G" Bamako-Mali

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***Corresponding author:** Traore Ousmane
Medical Imaging Department of the ''Marie Curie'' Medical Clinic in Bamako-Mali

Abstract **Case Report**

The sign of the scalp is the presence of perirenal fibrosis classically recognized as pathognomonic of Erdheim- Chester disease, a rare non-Langheransian histiocytosis. We report the case of a 31-year-old woman, wrongly followed and treated for bilateral polycystic kidney disease. She had no other medico-surgical history and had presented abdominal pain accentuated at the lumbar level, revealing on the abdomino-pelvic computed tomography performed at the "Marie Curie" Medical Clinic in Bamako, Mali, an aspect of hairy kidney. The diagnosis of non-Langheransian histiocytosis was retained in view of this specific and pathognomonic aspect. This was, to our knowledge, the first case described in Mali, the purpose of this work being to illustrate the role of CT in the diagnostic management of Erdheim- chester disease and to draw the attention of young radiologists and other specialists on the pathognomonic aspect of the scalp. The appearance of a scalp being considered semiologically as a very suggestive sign of Erdheim-Chester disease, the radiologist and his urologist or nephrologist counterparts must recognize these signs and finally not wrongly treat patients with this rare pathology.

Keywords: Scalp kidney, computed tomography, histiocytosis, Erdheim-Chester disease, incidental finding, Bamako.

INTRODUCTION

The sign of the scalp is the presence of perirenal fibrosis classically recognized as pathognomonic of Erdheim- Chester disease, a rare non-Langheransian histiocytosis [1]. It is a group L histiocytosis of the revised WHO classification of histiocytosis first described by Jakob Erdheim and William Chester in 1930 [2]. It is an inflammatory myeloid neoplasia which mainly affects male adults (3 men for one woman); rarely in children and the elderly [2]. This pathology is characterized by its multi-systemic attack which affects many organs with bone, retroperitoneal, pulmonary, cardiovascular, neurological and orbital localizations. The diagnosis is often made during the 5th decade with positive immunostaining for CD68 and negative for CD1a [3-5]. The pathophysiology of this condition is poorly understood. The prognosis depends on the affected organs [6-8]. Few data concerning the aspect in imagery in Mali and in the literature given the extreme rarity of the series. We report the case of a renal histiocytosis diagnosed here by a scanographic appearance of hairy kidneys in Bamako, the aim of which is to illustrate the role of the scanner in the diagnostic management of Erdheim-chester disease and to draw the attention of young radiologists and other specialists to the pathognomonic aspect of the scalp.

OBSERVATION

We report the case of a 31-year-old patient, wrongly followed and treated for bilateral polycystic kidney disease for more than 6 months. The patient had no other medical and surgical history. Biologically the renal function was good. Diffuse abdominal pain more

marked at the lumbar level had prompted the patient to consult again and an abdominopelvic CT scan was requested as part of the assessment. This imaging examination was carried out at the Medical Clinic " Marie Curie " in Commune V of the district of Bamako in Mali by GE brand equipment (general electric) of the OPTIMA 16 Barette scanner type. He had shown nephromegaly with bilateral perirenal fibrosis (resulting in the appearance of hairy kidneys) infiltrating the adrenal glands and in places the renal sinuses without significant hydronephrosis (Figures 1 and 2).

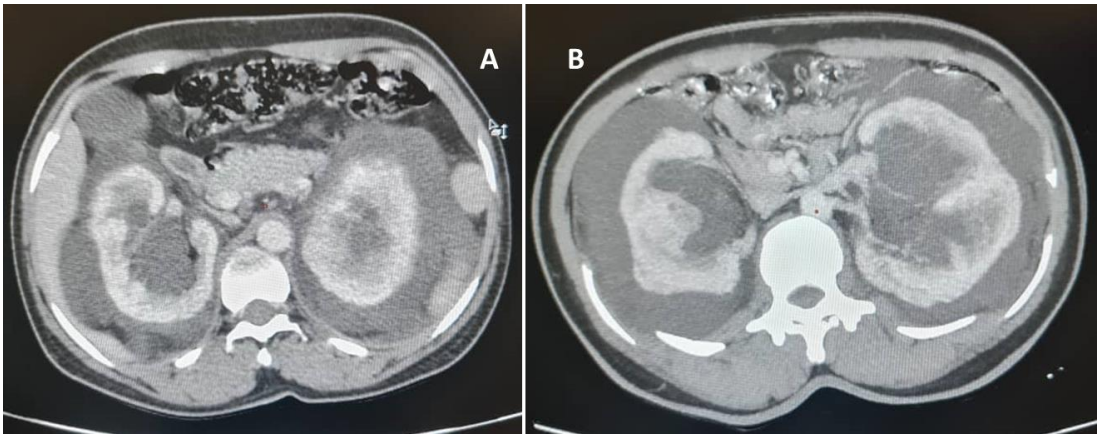

Figure 1 (A and B): Abdominal CT with axial reconstruction and MIP on both kidneys showing the appearance of hairy kidneys

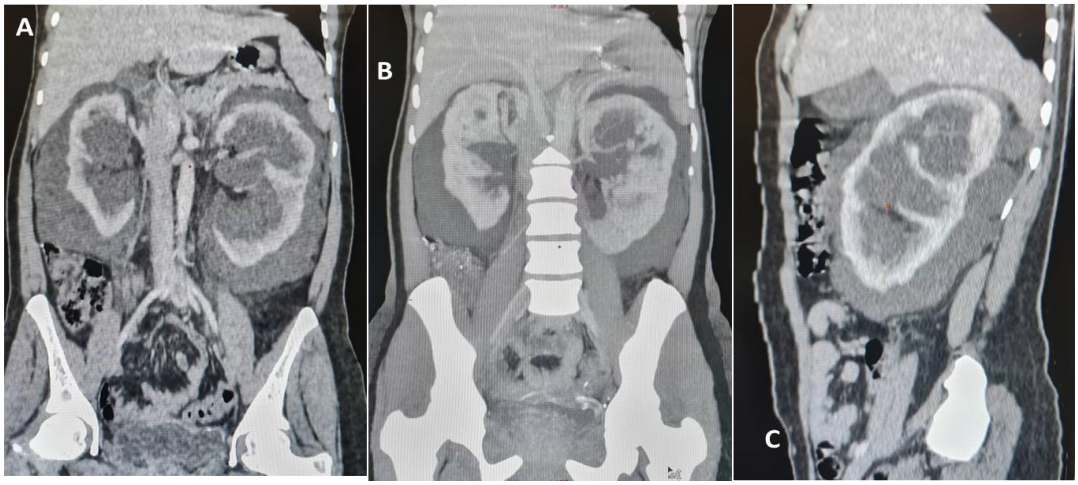

Figure 2 (A, B and C): Abdominal CT in coronal reconstruction (A and B) and sagittal (C) showing bilateral perirenal fibrosis giving an appearance of hairy kidneys

We also noted the presence of a few bilateral renal cysts at the cortical level of both kidneys (Figure 3).

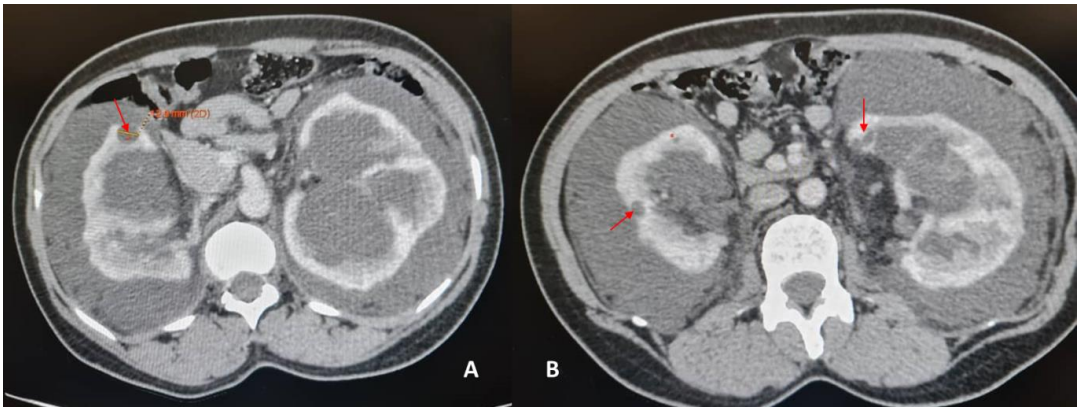

Figure 3(A and B): Abdominal CT with axial reconstruction with MIP passing through both kidneys showing small cortical cysts (red arrow)

The rest of the abdominal floor and the pelvis were normal on computed tomography. The bone window had not found any osteocondensing lesion that day. The diagnosis of non-Langheransian histiocytosis was retained in view of the specific and pathognomonic aspect of hairy kidneys and the diagnosis of polycystosis was abandoned despite the presence of 3 cysts per kidney. A biopsy was requested for histological confirmation. The patient was placed under clinical and radiological monitoring.

DISCUSSION

Erdheim Chester disease is a form of non-Langheransian, multi-systemic histiocytosis whose pathophysiology is poorly understood [6, 7]. It is rare and was discovered in 1930 by Jakob Erdheim and Williams Chester. The most frequent lesions are bone, renal, ocular, endocrine, neurological, and cardiovascular [7]. Our observation concerned bilateral renal involvement only. The predominance is male in 70% of cases and found in all ages [7, 8], our subject was female and 31 years old. The clinical signs depend on the organs affected, the asymptomatic forms exist and the systemic forms are aggressive, with a risk of life-threatening engagement in the event of damage to the noble organs [6, 7]. Bone manifestations are frequent and can represent 40% of cases according to Haroche *et al.*, [7]. In our case, the patient complained of diffuse abdominal pain and there was no bone involvement, only the kidneys were affected bilaterally. Imaging is characteristic with the presence of osteosclerosis and bone condensation on CT scan, and obvious hyperfixation on bone scintigraphy [4, 6, 7]. The long bone X-ray did not find any osteosclerotic lesion in our patient, on the scanner no bone lesion was also found. On the other hand, our patient had not benefited from a scintigraphy due to the lack of availability of this imaging in Mali. Involvement of the central nervous system and the orbit are described in the literature with a neurological clinical picture [9]. Our case did not present any neurological sign so she had not had an imaging of the head or the orbit (Magnetic Resonance Imaging I(RM)). On the basis of renal imaging, Erdheim-Chester disease is suspected, then confirmed by a biopsy of the renal capsule [8], it presents as retroperitoneal fibrosis described in 1/3 of cases often affecting the glands adrenal glands, the kidneys (appearance of hairy kidneys) and the excretory tracts with risk of obstructive renal insufficiency and Renovascular hypertension [4, 10, 11]. The typical imaging manifestations are retroperitoneal and perirenal fibrosis (hairy kidneys) often associated with periaortitis with circular sheathing (aortic sleeve) which can make the differential diagnosis with retroperitoneal fibrosis [12]. Computed tomography (CT) found in our patient a perirenal fibrosis with spiculated infiltration of the renal sinuses and the adrenal glands giving an appearance of hairy kidneys. There was no ureteral obstruction, therefore no hydronephrosis or periaortitis in our observation. The appearance on MRI is the same as on computed tomography [13]. Our patient could not benefit from an MRI given the difficult accessibility in Mali. There may also be cardiac involvement in the form of cardiomegaly, pericarditis up to tamponade, pleuropulmonary abnormalities in the form of inertial pneumonitis and pleurisy [6]. The biopsy of the affected organ with anatomopathological study confirms the diagnosis by showing histiocytic proliferation within fibrosis made of xantogranuloma with a vacuolated aspect of the cells. In our case, the biopsy could not be performed and we want to draw the attention of radiologists and other specialists to the appearance of the scalp bearing on the spiculated thickening of the renal capsule encountered in 68% of cases and pathognomonic of Erdheim Chester disease [5]. Our 31-year-old patient was wrongly treated for polycystic kidney disease given the presence of a few renal cysts despite the presence of the characteristic aspect of the scalp. The lack of experience on the pathognomonic aspect of perirenal fibrosis may explain this diagnostic error. There is no therapeutic consensus concerning the management of Erdheim Chester disease in the literature given the scarcity of series. The asymptomatic forms are subject to simple surveillance [6], our patient had been under surveillance for more than a month with a favorable clinical evolution.

CONCLUSION

The appearance of a scalp being considered semiologically as a very evocative sign of Erdheim-Chester disease, radiologists and their urologist or nephrologist counterparts must recognize these signs and finally not to wrongly treat patients with this rare pathology. CT is a reliable examination giving the characteristic appearance of the scalp in renal histiocytosis or Erdheim-Chester disease. Confirmation always remains histological.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

CONSENT

We obtained informed consent from the patient to use the CT image in this case report

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