

Exceptional Case

Extramedullary haematopoiesis in the kidney

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Abstract

Extramedullary haematopoiesis (EMH) is the development of haematopoietic tissue outside the bone marrow and it most often occurs in the liver and spleen. Renal EMH is quite rare and there are very few case reports concerning the kidney. We describe two cases of 'renal histologically documented EMH' and, in particular, in the second of these two, the EMH tissue coexists with a clear cell renal carcinoma. Although rare, these clinical pictures raise some questions about the role of needle biopsy in the management of renal masses that present a diagnostic dilemma, especially in cases without involvement of other abdominal or intrathoracic organs.

Keywords: extramedullary haematopoiesis; fine needle biopsy; kidney; renal mass

Introduction

Renal masses of uncertain aetiology may be discovered incidentally and in some cases, the diagnosis is difficult to establish based only on imaging examination. Extramedullary haematopoiesis (EMH) refers to the development of haematopoietic tissue outside the bone marrow and normally occurs in the reticuloendothelial system (liver, spleen and lymph nodes). The involvement of other parenchymatous organs is rare and there are only sporadic reports concerning the kidney [1–7, 8]. We describe two cases of 'renal histologically documented EMH', the first of which mimicked a bilateral malignant tumour of the kidney in a patient with a known history of polycythaemia vera, and the second was observed in an elderly male with a recent diagnosis of idiopathic myelofibrosis.

Case 1

An 80-year-old man, diagnosed with myeloproliferative disease (polycythaemia vera), was admitted after ultrasonography and computed tomography (CT) scan detection (Figure 1A) of bilateral parapyelic solid renal lesions that can simulate renal carcinoma. The right mass (6.5 cm sized) infiltrated the pelvicalyceal system, causing extrinsic mass effect and continued in the perirenal spaces. The left solid lesion was 2.3 cm in size. Investigations showed a haemoglobin level of 121 g/L (12.1 g/dL); white blood cell (WBC) $20.1 \times 10^9/L$ ($20.1 \times 10^3/\mu L$), platelet count (PLT) $82 \times 10^9/L$ ($82 \times 10^3/\mu L$); plasma creatinine 141.4 $\mu\text{mol/L}$ (1.6 mg/dL) and estimated glomerular filtration rate (eGFR) 0.70 mL/s (42 mL/min). As some doubts persisted about the moderate contrast enhancement, a CT-guided needle biopsy was performed without complications. The histological examination was compatible with the final diagnosis of

EMH, containing cells of three distinct lineages including myeloid and erythroid cells and rare megakaryocytes. The immunohistochemical staining was positive for myeloperoxidases and glycophorin (Figure 2), while CD34 staining was negative. No other signs of EMH were detected in the abdominal parenchymas. The conclusion of a subsequent bone marrow biopsy indicate myelofibrosis post-polycythaemia and he was treated with hydroxyurea and allopurinol. The patient is still alive 28 months after hospital admission.

Case 2

A 79-year-old man with a previous history of ischaemic cardiopathy was admitted to another department with persistent fever and complaints of fatigue and weakness. Examination revealed splenomegaly. Haemoglobin was 113 g/L (11.3 g/dL), WBC $17 \times 10^9/L$ ($17 \times 10^3/\mu L$), PLT $676 \times 10^9/L$ ($676 \times 10^3/\mu L$); plasma creatinine 101.6 $\mu\text{mol/L}$ (1.15 mg/dL); eGFR 1.01 mL/s (61 mL/min) and lactate dehydrogenase 1394U/L. Abdominal ultrasonography and magnetic resonance imaging (MRI) showed a perirenal infiltrating tissue associated with hepatosplenomegaly. Given the suspicion of lymphoma, the patient performed an osteomedullary biopsy that showed idiopathic myelofibrosis. A CT (Figure 1B) confirmed bilateral perirenal tissue with modest contrastographic impregnation and showed a solid mass in the lower pole of the right kidney with intense contrast enhancement. The patient was referred to us for a CT-guided needle biopsy that revealed the co-presence of two different lesions to the right a clear cell renal carcinoma, while the bilateral perirenal tissue was haematopoietic tissue, confirmed by immunohistochemical cell phenotype. The patient underwent a polar right nephrectomy and he is still alive 24 months after diagnosis with a minor renal dysfunction,

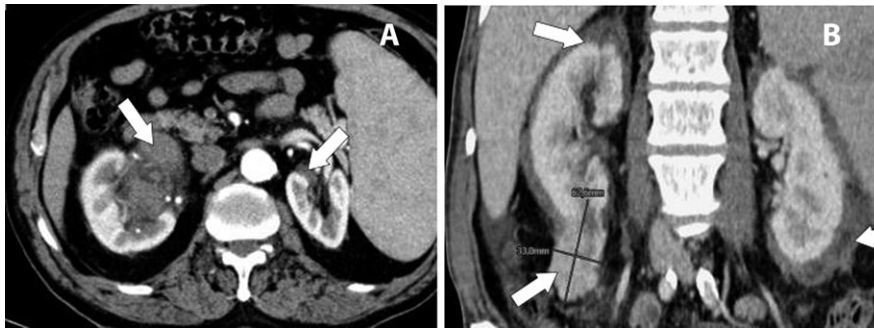


Fig. 1. (A) CT scan of two solid renal lesions with moderate contrast enhancement, one to the right and one to the left, in the parapelvic site, that can simulate a renal carcinoma (Case 1). (B) CT scan of perirenal infiltrating tissue (specimen confirmed at a subsequent MRI) giving the suspicion of lymphoma (Case 2).

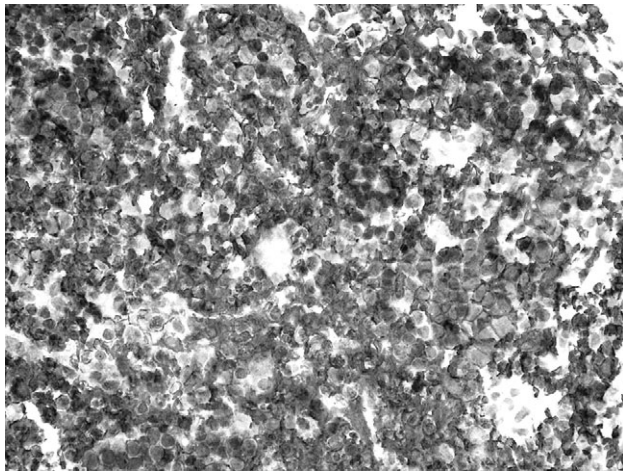


Fig. 2. Fine needle renal biopsy of Patient 1; erythroid cells positive for glycophorin staining.

plasma creatinine 114.9 $\mu\text{mol/L}$ (1.3 mg/dL) and eGFR 0.88 mL/s (53 mL/min).

Discussion

The kidney is an unusual site for the occurrence of EMH, and clinically, renal EMH can be asymptomatic. There have been <20 previous reports of EMH renal involvement [1–7, 8]. Renal involvement can be parenchymal, intrapelvic or perirenal. In the parenchymal type, the kidneys may either be enlarged or have focal lesions and the masses may be indistinguishable from renal cell carcinoma [2, 3]. Pelvicalyceal or hilar involvement is often an extension of a parenchymal lesion pattern and in this site, the EMH tissue may cause obstructive renal failure [4, 5]. In the perirenal type, the soft tissue encases both kidneys, such as in our Case 2. The bilateral perirenal localization of EMH may sometimes mimic a renal lymphoma [6].

The differential diagnosis of a perirenal or parapelvic mass of uncertain aetiology includes tumours, lymphomas, lipomatosis and renal inflammatory or infectious tissue [2, 6]. The role of nuclear medicine imaging or of FDG-PET/CT, to resolve such diagnostic problems, is still a controversial issue [6]. In both our cases, we chose a CT-guided biopsy approach to arrive at a final diagnosis. A histologically proven diagnosis of EMH in our patients could avoid unnecessary nephrectomy and contribute to preserve their renal function.

In all but two previous reports, renal EMH has occurred in association with chronic haematological disorders. In two cases, small foci of ‘pure erythropoiesis’ were found in areas within a clear cell renal carcinoma and in patients without underlying haematological disease [9, 10]. In both of these cases, the authors suggested that the abnormal erythroid proliferation may have been related to a local erythropoietin (EPO) excess produced by malignant cells [9, 10]. On the contrary, our Case 2 is quite unique because we found the coexistence of a lower pole clear cell renal carcinoma and perirenal EMH encasing both kidneys in a patient with concomitant idiopathic myelofibrosis. We believe that in our second case, the EMH was due to haematological disease rather than to an EPO excess.

The pathophysiology of solid organ involvement in EMH is still not fully understood. It has been speculated that haematopoietic cells are derived from resident mesenchymal pluripotent cells that could proliferate as a response to a disease-related simulating factor or may arise from migration of stem cells from bone marrow [8]. The renal localization presents some intriguing aspects: (i) does the kidney, with a scarcely represented reticuloendothelial tissue, maintain in adult life a niche for haematopoietic stem cell differentiation? (ii) Is a local intrarenal EPO excess able to drive stem cell migration and to promote EMH proliferation?

Conclusions

EMH in the kidney represents an interesting ‘speculative challenge’ in terms of differential diagnosis with other soft tissue masses. Guided ultrasound or guided CT needle renal biopsy might be included in the diagnostic algorithm to better manage the dubious cases and it also may be very useful to guide a less aggressive treatment.

Conflict of interest statement. None declared.

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