CASE REPORT

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An Unusual Case of Renal Extramedullary Haematopoiesis

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Abstract

Background: Extramedullary haematopoiesis, the proliferation of haematopoietic cells is an entity occurring in conditions with insufficient haematopoiesis.

Case Presentation: We report a rare case of renal extramedullary haematopoiesis (EMH) presenting as mildly bulky kidneys and a renal hilar and pelvic mass mimicking urothelial malignancy in a patient with pancytopaenia on haemogram. There was no osteosclerosis, paravertebral masses, or massive splenomegaly, the usual radiological features of a myeloproliferative disorder on imaging.

Conclusion: The possibility of renal EMH should be considered, in the presence of bulky kidneys or a renal pelvic mass not only in the background of the classical imaging clues, but even without the telltale signs of a myeloproliferative disorder on imaging if the blood picture suggests so.

Key words: Renal mass, extramedullary, haematopoiesis.

Introduction

Extramedullary haematopoiesis (EMH), rarely encountered in general radiologic practice, is a common feature of chronic myeloproliferative disorders. EMH occurs in response to inadequate erythropoiesis in the bone marrow. If red marrow reconversion is unable to meet the body's demand for blood cells, haematopoiesis will shift outside of the bone marrow, in most cases occurring in the paravertebral region or the liver and spleen, the site of physiological EMH during fetal life[1,2]. Rarely, other organs can be involved. Renal involvement by EMH is relatively uncommon and can present in various forms like parenchymal, pelvic, or perirenal.

Herein we report a rare case of renal EMH in a patient with pancytopaenia with the absence of osteosclerosis and other radiological signs of a myeloproliferative disorder on imaging.

Case Report

A 50-year-old male presented with complaints of chronic fatigue and weakness. There was no history of fever or haematuria. A haemogram revealed pancytopaenia. Ultrasonography (USG) of the abdomen showed mild hepatosplenomegaly, mildly bulky kidneys and a hyperechoic mass in the left renal sinus conferring a faceless appearance to the left kidney.

Multiphasic Computed tomography (CT) of the abdomen revealed a rounded contour of both kidneys with a homogenous, hypo-attenuating, and minimally enhancing soft tissue density mass replacing the left renal sinus and extending into the left renal hilum, obscuring the renal vessels (Figs. 1a, 1b, 2a). The soft tissue density was seen to fill and expand the renal pelvis and proximal ureter (Fig. 1a, 2b). Few tiny (5 to 7 mm) round, focal hypoenhancing lesions were seen scattered in the bilateral renal parenchyma.

There were no osteosclerotic or paravertebral soft tissue masses. Primary urothelial mass and lymphoma were considered as possibilities and a USG-guided Fine Needle Aspiration Cytology (FNAC) was carried. The bone marrow

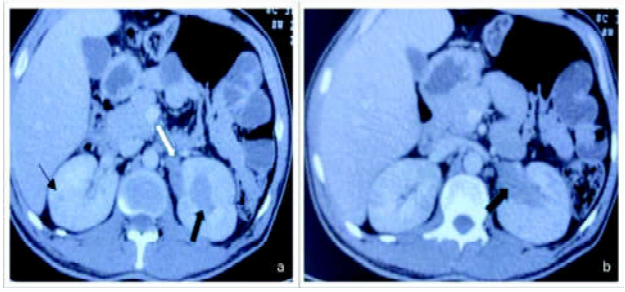

Fig. 1a: *Axial CECT image shows rounded contour of both kidneys with multiple small, round, focal hypoenhancing lesions (thin black arrow) scattered in bilateral renal parenchyma. A soft tissue density mass is seen replacing the fat in the left renal sinus, (black arrow) and encasing and filling the left renal pelvis (white arrow).*

Fig. 1b: *Axial CECT image shows expansile hypoenhancing soft tissue density mass (black arrow) in left renal hilum.*

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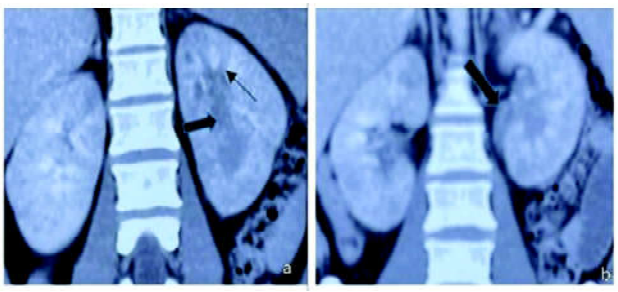

Fig. 2a: *Coronal CECT image shows shows hypoenhancing soft tissue density mass (thick black arrow) in the left hilum stretching the calyces (thin black arrow).*

Fig. 2b: *Coronal CECT (3 mm MIP) image shows hypoenhancing soft tissue density mass (black arrow) in the left renal hilum filling and expanding the left renal pelvis and proximal ureter.*

biopsy, given pancytopaenia, revealed marrow fibrosis with a paucity of normal haemopoietic elements consistent with myelofibrosis. Fine needle Aspiration cytology (FNAC) from the left renal pelvic mass revealed a polymorphous infiltrate composed of immature erythroid cells, myeloid cells, fat cells, and lymphocytes (Fig. 3), consistent with extramedullary haematopoiesis.

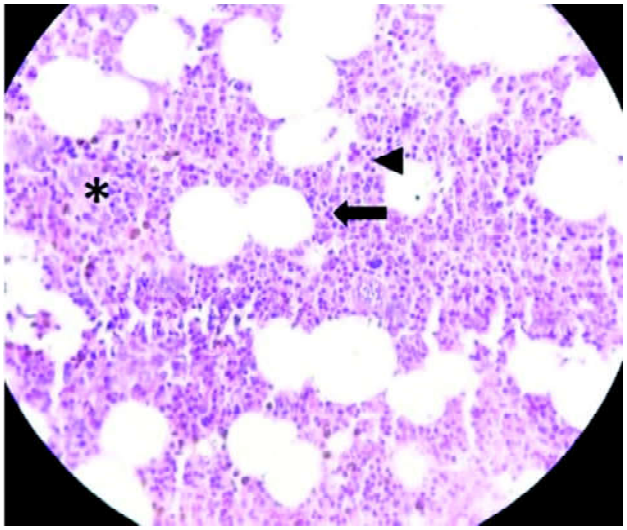

Fig. 3: *40 X view of haematoxylin and eosin (H & amp; E) stained section showing aggregates of haematopoeitic precursor cells including erythroblasts (arrowhead), myeloid cells (arrow), fat cells (thin arrow) megakaryocytes* within the parenchyma.*

Discussion

Extramedullary haematopoiesis (EMH) is a compensatory mechanism for inadequate haemopoiesis. It refers to deposits of erythroid precursors in sites other than the bone marrow.

The haematopoietic and stromal cell lines occur in bone marrow and organs like the spleen and liver which were haematopoietic in the fetus. In conditions such as myelofibrosis with myeloid metaplasia, control of stem cell differentiation is lost, and peripheral blood cytopaenia occurs due to haematopoietic elements of the marrow being replaced by fibrosis.

Neoplastic stem cells can circulate and migrate to secondary haematopoietic organs giving rise to EMH, particularly in the spleen, liver, and lymph nodes that are part of the reticuloendothelial system. Even though the reticuloendothelial system is the main site of EMH, other organs such as the lungs, gastrointestinal tract, breast, skin, kidneys, and adrenals can be recruited for haemopoiesis either due to presence of haematopoietic precursors or due to the circulating stem cells being deposited in these organs[1,2,3].

The differential diagnosis of a renal pelvic mass on CT includes lymphoma, urothelial mass, and renal sinus lipomatosis[3]. Lipomatosis typically has very low attenuation and does not enhance on CT. Renal lymphoma may present as either a mildly enhancing perirenal mass encasing the entire a kidney or renal hilum and pelvicalyceal system[4]. Intrapelvic transitional cell carcinoma typically manifests as a hypoenhancing intrapelvic mass with expansile growth where the shape of the kidney remains intact.

Renal involvement by EMH can present as a hypoattenuating and hypoenhancing mass in the perirenal, parenchymal, intra or para-pelvic location. In parenchymal type of involvement, the kidneys may either be enlarged diffusely or have small focal lesions. In perirenal type of involvement, a hypoattenuating, hypoenhancing uniform mass or nodules are seen engulfing the kidneys without distorting their shape. Pelvic involvement could be an extension of parenchymal lesions, but can also be isolated. In our case there was a urothelial mass involving left renal pelvis and proximal ureter extending to the renal sinus encasing the pelvicalyces.

Additionaly, co-existing parenchymal involvement of both kidneys was seen on imaging as rounded contour of kidneys with small focal hypoenhancing lesions on CT. Bulky kidneys can also be seen in lymphoproliferative disorders.

Clinically, renal EMH can be asymptomatic or can present with symptoms ranging from abdominal discomfort to renal failure due to either ureteral obstruction or extensive parenchymal involvement resulting in interstitial nephritis[5].

Most reports have mentioned renal extramedullary haematopoiesis presenting on imaging as a homogeneous, hypoattenuating, soft tissue mass showing minimal enhancement on CT as in our case. In most of these reports, bilateral renal masses encasing the pelvicalyces were present[3,5,6]. EMH presented as solitary renal parenchymal

mass in the reports by Ahuja *et al*[8] and Mubeen *et al*[9]. Peri renal mass engulfing renal parenchyma was seen in the case report by Ricci *et al* and Imai *et al*[7,10].

Kurien *et al* reported a case of EMH presenting as acute renal injury due to parenchymal involvement causing interstitial nephritis[5]. In our case there was parenchymal involvement causing bulky kidneys and tiny lesions in bilateral renal parenchyma. All these case reports have shown radiological features of an underlying myeloproliferative disorder like massive splenomegaly and osteosclerosis on imaging.

In the right clinical setting, CT can indicate a diagnosis of EMH. Our case was unique as there were no osteosclerosis, thoracic paravertebral masses, or substantial splenomegaly to suspect EMH in the presence of myelofibrosis. Our case was further challenging as there was a unilateral mass in the renal pelves with features overlapping with a transitional carcinoma involving the pelvis and proximal ureter.

Conclusion

Despite many reports, little attention has been paid to the overall spectrum of imaging findings of renal extramedullary haematopoiesis. EMH should not only be included in the differential diagnosis of a non-specific renal mass in a confirmed case of myelofibrosis but should also be considered in an incidental renal mass in cases with peripheral pancytopaenia lacking radiological features of an underlying myeloproliferative disorder.

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