Primary Ewing’s Sarcoma of the left kidney

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Section: Uroradiology & genital male imaging
Area of Interest: Abdomen Kidney
Procedure: Diagnostic procedure
Imaging Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 26 years, male

Clinical History:

A 26-year-old male patient presented with left lower quadrant pain. Initial computed tomography (CT) of the abdomen and pelvis was performed to rule out diverticulitis. Follow up CT of the abdomen and pelvis was performed to further delineate the left renal mass found on initial imaging.

Imaging Findings:

Initial CT of the abdomen and pelvis showed an irregular, hypodense, mildly exophytic mass within the lower pole of the left kidney measuring 3.3 cm (AP diameter), 2.9 cm (craniocaudal) and 2.9 cm (transverse diameter). No hydronephrosis was appreciated and the left renal vein showed no filling defects. Aorta and IVC were normal. No lymphadenopathy was appreciated. Adrenals were normal (Fig. 1).

Secondary imaging showed the mass to have an inhomogeneous pattern of contrast enhancement. Areas of lower density were within the medial aspect of the lesion suggestive of necrosis. No calcifications were identified within the lesion. The lesion involved both the cortex as well as the medulla of the kidney and extended into the region of the renal sinus (Fig. 2). Obliteration of the perisinus fat as well as displacement of the calyces was noted. There was no evidence of distant metastases.

Discussion:

Extraskeletal Ewing’s Sarcoma (EES) was first described in 1975 and is part of the Ewing’s sarcoma family of tumours (ESFT). ESFT represents a group of round cell tumours and includes peripheral primitive neuroectodermal tumour (PNET), thoracopulmonary PNET (Askin tumor), skeletal Ewing’s sarcoma, and EES [1].

EES is a rare diagnosis, but has a male preponderance of 1.6:1, and most often occurs in patients between 3 and 25 years of age (mean: 13 years) [2]. Primary Ewing’s sarcoma is an aggressive neoplasm with very poor prognosis. The 5-year disease free survival rate is estimated to be 45-55%, and 25-50% of patients first present with metastatic disease [3]. Although it is exceedingly rare, a limited number of cases have shown EES infiltration of the kidney [4]. Due to its rarity, this is often mistaken for a number of other round cell tumours such as Wilms tumour, monophasic synovial sarcoma, lymphoma, clear cell sarcoma of the kidney, neuroblastoma, and most commonly, renal cell carcinoma (RCC) [3]. It is important to note that primary Ewing’s arising of the kidney appears to behave even more aggressively than EES at other sites [3].

Renal cell carcinoma accounts for 90% of all adult renal cell primary malignancies [5]. On contrast-enhancement,
RCC is often solid with areas of hypodensity suggestive of necrosis. Given the similar features of RCC to our lesion, this was initially thought to be in keeping with RCC. At our institution, biopsies are not used to confirm RCC, thus nephrectomy was performed. Post-resection analysis of the kidney was required for a definitive diagnosis and found characteristic immunohistochemical and molecular markers of a primary Ewing’s sarcoma. To our knowledge, this is the first case of its kind at our institution.

While there is limited literature documenting the characteristic imaging features of renal Ewing’s sarcoma lesions, it appears that imaging is primarily utilized to rule out other neoplasms such as Wilms tumour. The presence of calcifications, tumour uptake of bone-seeking radiopharmaceuticals, and bone metastases are three main imaging features that help distinguish EES of the kidney from Wilms tumour [3]. Nevertheless, molecular and immunohistochemical analyses remain the mainstay for definitive diagnosis [4]. As more cases are documented, consideration of EES will become more prevalent when presented with a renal mass and this will help direct clinical suspicion and expedite definitive diagnosis. Increased awareness will further help in optimizing patient care and prognosis.

**Differential Diagnosis List:** Primary Ewing’s sarcoma of the left kidney, Renal cell carcinoma, Renal sarcoma, Metastatic disease, Haematologic malignancy (lymphoma), Wilms tumour

**Final Diagnosis:** Primary Ewing’s sarcoma of the left kidney.

**References:**


**Description:** Axial CT acquired in the venous phase showing an irregular, hypodense, mildly enhancing and slightly exophytic mass in the lower pole of the left kidney. **Origin:** Department of Radiology, Vancouver General Hospital, Vancouver, BC, Canada
Description: Coronal CT acquired in the venous phase showing an irregular, hypodense, mildly enhancing and slightly exophytic mass in the lower pole of the left kidney. Origin: Department of Radiology, Vancouver General Hospital, Vancouver, BC, Canada
Figure 2

Description: Axial CT acquired in the excretory phase further characterizing the known left renal mass showing inhomogenous contrast enhancement, areas suggestive of necrosis, and no identifiable calcifications. The lesion involves both the cortex and medulla. Origin: Department of Radiology, Vancouver General Hospital, Vancouver, BC, Canada
Description: Coronal CT acquired in the excretory phase further characterizing the known left renal mass showing inhomogenous contrast enhancement, areas suggestive of necrosis, and no identifiable calcifications. The lesion involves both the cortex and medulla. Origin: Department of Radiology, Vancouver General Hospital, Vancouver, BC, Canada