

CASE REPORT

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Ewing's sarcoma of the right kidney with multiple metastasis on PET/CT

Man Mohan Singh¹, Shashwat Verma^{1*} , Lavish Kakkar¹, Satyawati Deswal¹ and Nuzhat Husain²

Abstract

Background: Ewing's sarcoma is categorized as small round cell aggressive tumor which commonly occurs in bone and soft tissue. Primary renal Ewing's sarcoma is an extremely rare entity and accounts for less than 1% of renal masses. It is characterized by a very aggressive course, with very few reported cases in the literature. Primary renal Ewing's sarcoma is indistinguishable from renal cell carcinoma on anatomical imaging and therefore need histopathological correlation for confirmation. The positron emission tomography (PET) fused with computed tomography (CT) helps in the evaluation of disease, for staging and metastatic workup.

Case presentation: We present a case of 48-year-old lady who presented to urology department with difficulty in passing urine for 5 months associated with pain in the right flank region. She had an episode of haematuria 5 months back. Contrast enhanced CT showed a large lobulated heterogeneously enhancing right retroperitoneal mass in renal fossa. Histopathological examination and immunohistochemistry were suggestive of Ewing's sarcoma. The PET/CT revealed FDG avid right renal mass with lymph nodal and skeletal metastasis.

Conclusions: Ewing's sarcoma being aggressive tumour usually present with bone, liver and lung metastasis along with lymph nodal metastasis at time of diagnosis. The ¹⁸F FDG PET/CT helps in diagnosis, staging, appropriate treatment planning, restaging, response evaluation and prognostication of Ewing's sarcoma.

Keywords: Ewing's sarcoma, ¹⁸F FDG PET CT, Renal mass, Retroperitoneal mass

Background

Ewing's sarcoma is typically a bone or soft tissue, round cell sarcoma of primitive neuroectodermal origin. It rarely occurs as a primary renal neoplasm [1] and exhibits highly aggressive biological behaviour [2]. Fewer than 150 cases are reported till date [3] and most patients are young adults in the age group of 20 to 40 years with rare paediatric case reports. Primary Renal Ewing's sarcoma is highly malignant, grows rapidly, and metastasizes early to the lung, bone and lymph node. We present a case of 48-year-old female patient with primary renal Ewing's sarcoma who was referred for ¹⁸F FDG PET CT to our department of nuclear medicine for staging of disease.

Case presentation

A 48-year-old female presented to the urology department with difficulty in passing urine for 5 months associated with pain in the right flank region. She had an episode of haematuria 5 months back. The pain was intermittent in nature, with no acute exacerbations. She underwent contrast enhanced CT (CECT) abdomen which showed a large lobulated heterogeneously enhancing retroperitoneal mass lesion which had completely replaced the right kidney. It also showed multiple areas of internal necrosis and haemorrhage without any fat component or calcifications. The mass was extending superiorly with scalloping of the inferior surface of the right lobe of the liver with focal loss of fat planes. Medially it was crossing the midline, abutting the aorta more than,, renal vein, posteriorly extending up to the posterior abdominal wall and anteriorly abutting the antropyloric region of the stomach, duodenum and pancreatic head.

*Correspondence: drshashwat81@gmail.com

¹ Department of Nuclear Medicine, RMLIMS: Dr Ram Manohar Lohia Institute of Medical Sciences, Lucknow, Uttar Pradesh, India
Full list of author information is available at the end of the article

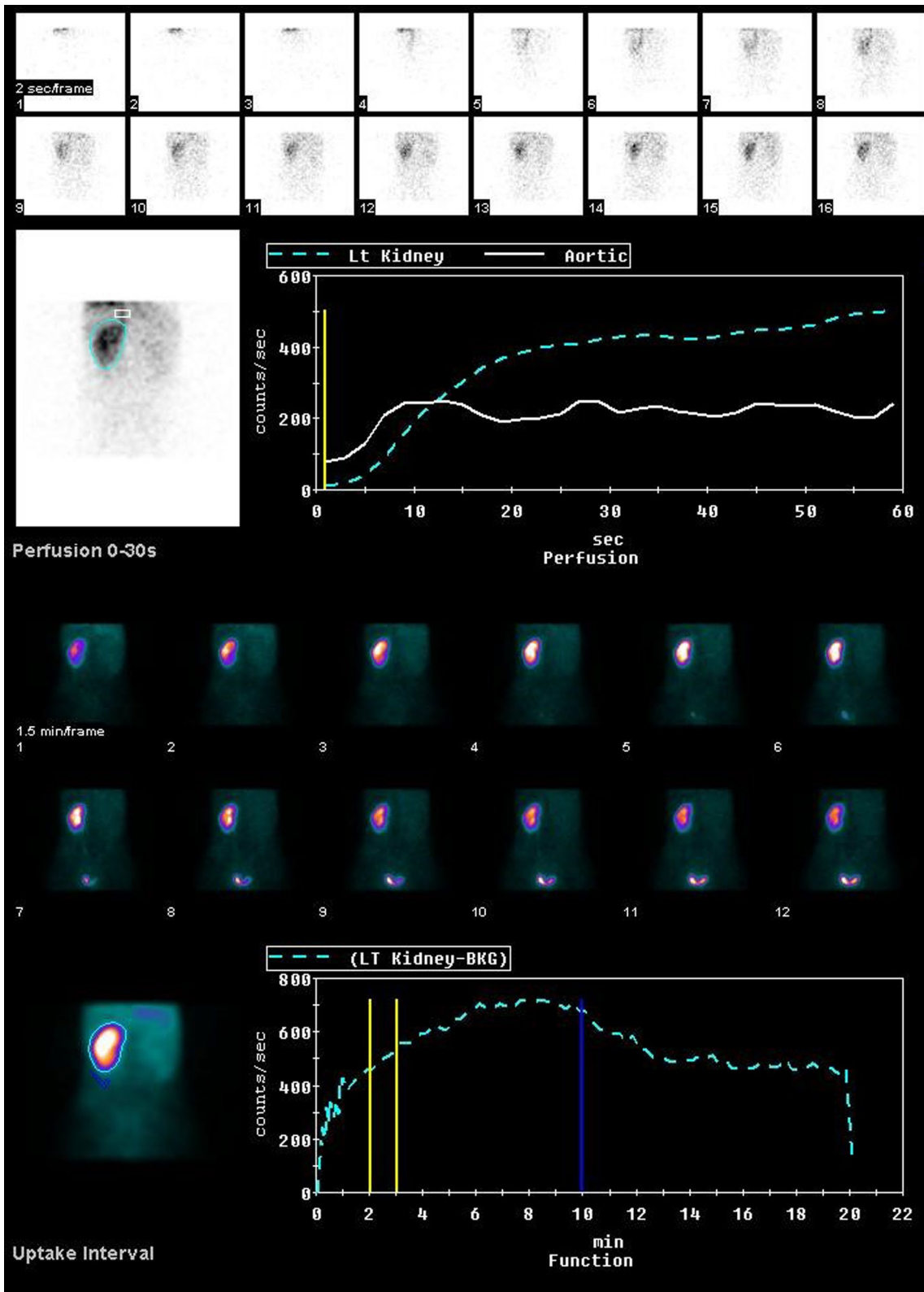


Fig. 1 99 m Tc- EC scan reveals non-visualization/ non-functioning right kidney

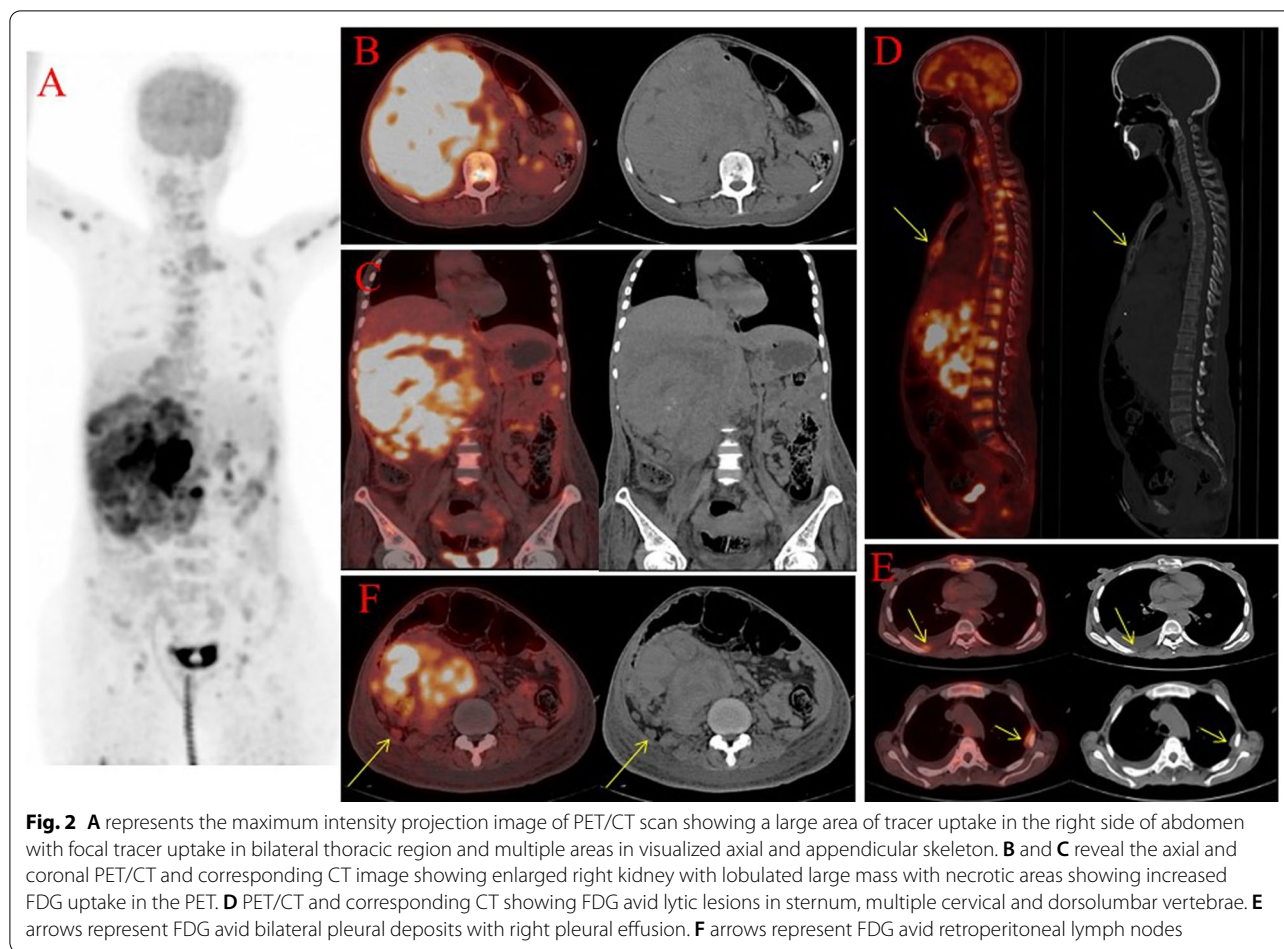


Fig. 2 **A** represents the maximum intensity projection image of PET/CT scan showing a large area of tracer uptake in the right side of abdomen with focal tracer uptake in bilateral thoracic region and multiple areas in visualized axial and appendicular skeleton. **B** and **C** reveal the axial and coronal PET/CT and corresponding CT image showing enlarged right kidney with lobulated large mass with necrotic areas showing increased FDG uptake in the PET. **D** PET/CT and corresponding CT showing FDG avid lytic lesions in sternum, multiple cervical and dorsolumbar vertebrae. **E** arrows represent FDG avid bilateral pleural deposits with right pleural effusion. **F** arrows represent FDG avid retroperitoneal lymph nodes

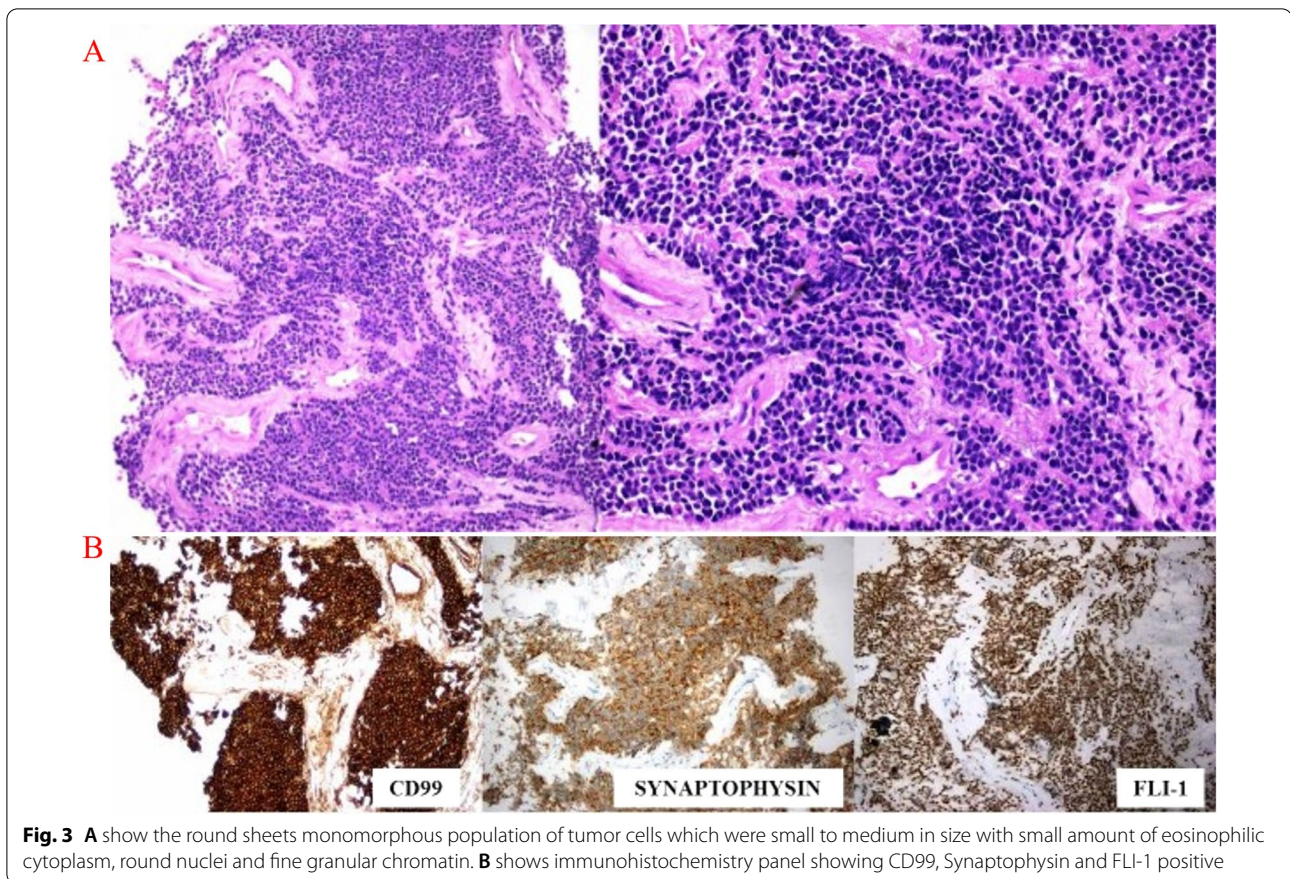
Multiple large veins, including the right renal vein, left common iliac and infrahepatic part of IVC show thrombosis. The findings were suggestive of renal cell carcinoma. The renal scan using ^{99m}Tc ethylene dicycstine (EC) was done to check the functional status of the right kidney and there was non-visualization of the right kidney during the study course and in delayed images suggestive of non-functional status of the kidney (Fig. 1). The patient was referred to our department of nuclear medicine for ^{18}F FDG PET CT for staging of disease. The ^{18}F FDG PET CT (Fig. 2) showed FDG avid ill-defined large mass ($\sim 14.8 \times 18.7 \times 20.6$ cm, SUVmax 17.6) arising from the right kidney abutting the liver with anterior displacement, medially it is crossing the midline abutting the aorta, corresponding renal vein superior mesenteric and inferior mesenteric artery, posteriorly extending up to posterior abdominal wall and anteriorly abutting antro-pylori region of stomach, duodenum and pancreatic head. Multiple FDG avid lytic lesions were noted in the sternum (\sim SUVmax 4.2), multiple ribs bilaterally, bilateral scapulae, multiple vertebrae, sacrum, bilateral pelvic bone, bilateral humeri and femora. FDG avid

discrete pleural deposits (\sim SUVmax 4.1) were noted in bilateral lungs. The histopathology of the renal mass was done and it showed sheets of monomorphous population of tumour cells which were small to medium in size with amounts of eosinophilic cytoplasm, round nuclei and fine granular chromatin. Immunohistochemistry markers including NFX-2.2, FLI-1, CD-99, Synaptophysin were positive (Fig. 3) and Ki 67 proliferation index was 70–80%. The histopathological findings were consistent with Ewing's sarcoma.

As the histopathological findings were suggestive of Ewing's Sarcoma, the patient's treatment changed from surgical management (as previously planned) to medical management. However, the patient succumbed to the disease before treatment could be initiated.

Discussion

Ewing's sarcoma rarely presents as primary kidney tumor [4]. The clinical features are usually non-specific such as flank or abdominal pain, hematuria or palpable mass. No specific features are seen in imaging and thus it is impossible to



differentiate renal Ewing's from much more prevalent RCC by imaging alone. RCC and sarcomas can have a similar heterogeneous appearance with areas of necrosis, hemorrhage, and calcification. [5]. Primary renal sarcomas are diagnosed on imaging only after exclusion of renal metastatic involvement from a primary sarcoma, secondary renal involvement from a retroperitoneal sarcoma, and ruling out sarcomatoid renal cell carcinoma. However, definitive diagnosis of renal Ewing's is solely by post- or pre-operative biopsy [6].

Ewing's sarcoma being the aggressive tumor with chances of early metastasis and can have bone, liver and lung metastasis along with lymph nodal metastasis at time of diagnosis [7].

^{18}F FDG PET/CT has shown to help to localize the primary sarcoma, its extension, metastasis specially the bony and pulmonary metastasis and to check the response to standard treatment available [8]. It also allows the detection of progression or regression and recurrence of disease prior to anatomic imaging [8, 9].

Conventional imaging can assess the local resectability of tumor, however for distant metastasis, conventional imaging (including CT, MRI etc.) can diagnose only 70% of the cases [10]. Thus PET/CT helps in

detection of disease involvement of tissue even before the anatomical changes occurs. The PET/CT has 63–8% sensitivity and 75–100% specificity for detecting metastasis and recurrence in renal tumors. These findings are similar for different variants of renal tumor. The patients with localized disease have a 5-year disease free survival rate of about 45–55% [11]; however the prognosis is even worse in patients with metastatic disease, with overall cure rate of 20% only. The main challenge is the proper diagnosis and timely management.

Conclusions

Primary renal Ewing's sarcoma is rare disease in adults which can be kept as primary differentials. Because of non-specific clinical features, it is difficult to differentiate Ewing's sarcoma from RCC on imaging. Patients usually present with bone, liver and lung metastasis along with lymph nodal metastasis at time of diagnosis. The final diagnosis is made by histopathology and immunohistochemistry. The ^{18}F FDG-PET/CT can be used for evaluating the primary, metastatic disease, and in evaluating response to the chemotherapy.

Abbreviations

RCC: Renal cell carcinoma; CECT: Contrast enhanced computer tomography; FDG: Fluorodeoxyglucose; PET/CTP: Positron emission tomography-computed tomography.

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Author contributions

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Availability of data and materials

The dataset used and/or analysed during the current study available from the corresponding author on the reasonable request.

Declarations**Ethics approval and consent to participate**

Written consent to participate.

Consent for publication

Written consent for publication from study participant.

Competing interests

The authors declare that they have no competing interests.

Author details

¹Department of Nuclear Medicine, RMLIMS: Dr Ram Manohar Lohia Institute of Medical Sciences, Lucknow, Uttar Pradesh, India. ²Department of Pathology, RMLIMS: Dr Ram Manohar Lohia Institute of Medical Sciences, Lucknow, India.

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