Primary Small Cell Carcinoma of the Kidney: A Case Study with Emphasis on Fluorodeoxyglucose Positron Emission Tomography–Computed Tomography Findings

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Abstract

Small cell carcinoma (SCC) of the kidney is included in extrapulmonary SCC which is a group of extremely rare but highly aggressive cancers. There have been only a few case reports and small retrospective series in the literature describing the malignancy in kidneys. Most of the published reports describe the entity as a variant mixed with other tumor subtypes such as urothelial carcinoma, adenocarcinoma, and squamous cell carcinoma. Pure-form SCC in kidneys is exceedingly rare. Fluorodeoxyglucose positron emission tomography–computed tomography plays an essential role in the accurate staging evaluation of this cancer.

Keywords: *Fluorodeoxyglucose positron emission tomography–computed tomography, high-grade renal NET, renal NET, renal small cell carcinoma*

Introduction

Small cell neuroendocrine carcinoma or small cell carcinoma (SCC) of the kidney is one of the extremely rare forms of extrapulmonary SCC. The absence of native neuroendocrine cells in normal renal parenchyma is the proposed reason behind its rare occurrence. SCC in kidneys is theorized to arise from totipotent stem cells, developing *de novo* or else from well-differentiated neuroendocrine tumors, giving rise to mixed histology.[1-4]

Early and extensive metastases are a hallmark of the aggressive natural history of the disease.[5] While patients with locoregional disease may be salvageable with early and vigorous therapy, relapses are common and the overall prognosis is poor, with the median survival rate being around 8 months.[5] The final diagnosis is based on histopathology and immunohistochemistry (IHC). Fluorine-18-fluorodeoxyglucose (FDG) positron emission tomography–computed tomography (PET-CT) is a valuable tool for evaluating SCC, owing to poor histological differentiation of these tumors, unlike the well-differentiated neuroendocrine tumors.[6,7]

In this case report, we describe the spectrum of the disease extent in a case of SCC of the kidney on FDG PET-CT. To the best of our knowledge, this is the first report in the literature describing the FDG PET-CT imaging findings in a case of SCC kidney.

Case Report

A 50-year-old male patient presented with complaints of abdominal pain and fullness for 1½ months. Ultrasound abdomen and contrast-enhanced CT abdomen revealed an exophytic lesion in the upper pole of the left kidney along with multiple space-occupying lesions in the liver. The patient was presumed to be a case of renal cell carcinoma (RCC) and referred for FDG PET-CT for baseline staging. PET-CT demonstrated increased FDG uptake in a mass lesion arising from the lateral cortex of the upper pole of the left kidney (size 3.7 cm, SUVmax: 10.0) with multiple FDG-avid abdominal, pelvic, retrocrural, mediastinal, and bilateral lower cervical lymph nodes (largest 4.6 cm, SUVmax: 16.7), multiple FDG-avid lesions in the liver, bilateral adrenals, bilateral lungs, multiple bones, and a FDG-avid subcutaneous nodule in the anterior abdominal wall [Figure 1].

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Biopsy from one of the liver lesions was performed and histopathological examination showed a core of liver tissue infiltrated by a poorly differentiated carcinoma with solid-nested architecture. The IHC analysis revealed positivity for pancytokeratin, synaptophysin, and chromogranin with high Ki-67 proliferation index (75%–80%) indicating a diagnosis of SCC [Figure 2].

The patient was started on etoposide- and platinum-based combination chemotherapy in view of widespread metastases on the FDG PET-CT and has received two cycles of therapy with subjective clinical benefit and minimal chemotoxicity till the time of writing this report.

Discussion

Extrapulmonary SCCs are uncommon and highly aggressive neoplasms. Most of the SCCs involving the genitourinary tract arise from the urinary bladder and primary SCC in kidneys is rare, with only around

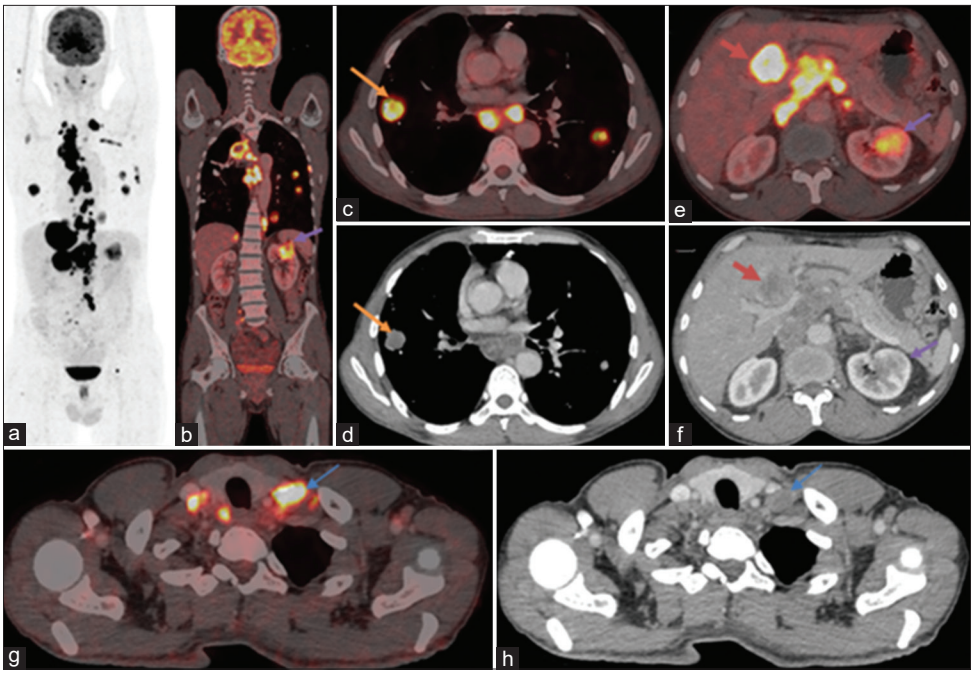

Figure 1: Fluorodeoxyglucose positron emission tomography–computed tomography (FDG PET-CT) images. Maximum intensity projection: (a) Fused PET-CT images coronal (b) and axial sections (c-h), showing a FDG-avid exophytic mass lesion arising from the lateral aspect of the upper pole of the left kidney (purple arrow, b, e and f), with multiple FDG-avid retroperitoneal, abdominopelvic, mediastinal, and bilateral cervical level IV and bilateral supraclavicular (blue arrow, g and h) lymph nodes, bilateral lung nodules (orange arrow, c and d), and lesion in the liver (red arrow, e and f)

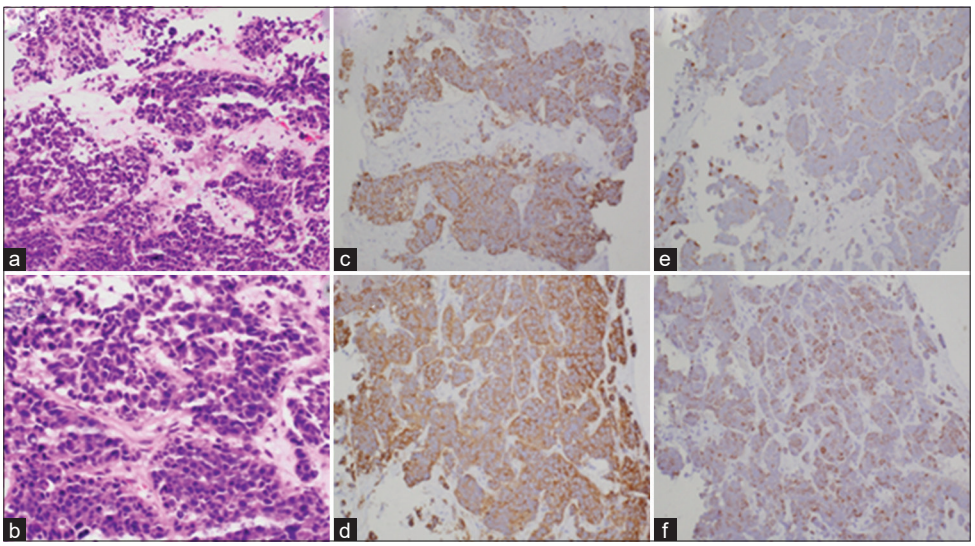

Figure 2: (a) Biopsy from liver space-occupying lesion shows tumor cells arranged in solid sheets with focal necrosis, H and E × 200. (b) The tumor cells display a high nucleocytoplasmic ratio, markedly pleomorphic hyperchromatic nuclei with molding, and inconspicuous nucleoli along with apoptoses and mitoses, H and E × 400. These cells are immunopositive for pancytokeratin (c), synaptophysin (d), chromogranin (e) and show a high Ki67 proliferation index (f), Immunohistochemistry × 200

60 cases having been described in the literature to date.[1,2] Women are more likely to be affected than men, with a median age at diagnosis of around 60 years.[5] There are no symptoms or clinical features pathognomonic for its diagnosis since the usual presenting complaints including abdominal pain and hematuria overlap with those of other renal malignancies. The diagnosis is confirmed after light microscopy and IHC analysis of the tumor sample. Limited disease is defined as a tumor restricted to the organ of origin and regional lymph nodes which can be included within one radiation treatment portal, whereas the presence of nonregional lymph nodes and involvement of distant visceral organs is regarded as extensive disease. Based on this classification, Majhail *et al*.[5] examined the literature and discovered that around two-thirds of reported patients had limited-stage illness. Our patient presented with extensive disease with multiple liver, lung, and nonregional lymph nodal metastases. A clinical classification system comparable to that used for SCC of the lung can be applied to SCC of the kidney.[6,8] SCC of the kidney can be further divided into subcategories of pure or mixed types. They can also be associated with urothelial carcinomas and less commonly develop in isolation.[9] In a retrospective observational study done by Masuda *et al*., 18 cases were described and more than half of the cases were associated with transitional cell carcinoma.[10] Our patient showed isolated SCC of the kidney with no association with other histologic subtypes.

Due to the aggressive clinical course and rarity of presentation, there are currently no established guidelines available for the management of this entity. Depending on the primary site and severity of the disease, multimodal therapy usually consists of radiation and chemotherapy in addition to potential surgery. The chemotherapy regimens utilized to treat extrapulmonary SCC are similar to those used for the lung origin SCC. Platinum-based chemotherapy is considered an acceptable adjuvant treatment after surgery aimed to lower the risk of systemic recurrence.[11] The most accurate indicator of survival is the disease severity at diagnosis with extensive disease having a very aggressive course and <1-year survival after diagnosis.[12,13]

Lee *et al*. observed the correlation between the SUVmax values of primary RCC with the presence of distant visceral metastases in 23 patients.[14] More than two-thirds of the patients had distant metastases, for whom the median SUVmax of primary RCC was 5.0 (range 2.9–7.6), and for the rest one-third with only locoregional disease, SUVmax was 2.6 (range: 1–5.6). In our case, the SUVmax value of the primary kidney tumor was 10.0, significantly above the values observed in high-grade RCC. The SUVmax values for metastatic lesions (liver, lung, and lymph nodes) were even higher, ranging between 11.3 and 17.3.

Conclusion

FDG PET-CT is useful in the staging of SCC of the kidney and helps in the determination of an appropriate course of treatment. The presence of renal mass on FDG PET-CT with a relatively higher SUVmax value should raise a suspicion for more aggressive renal tumors other than RCC, including lymphoma and SCC.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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