Case Report

Primary extrarenal papillary renal cell carcinoma presenting as a neck mass

S. Srivishnu, Ganesh Bakshi*, Santosh Menon[1]

Departments of Surgical Oncology and [1]Pathology, Homi Bhabha National Institute, Tata Memorial Hospital, Mumbai, Maharashtra, India

*E-mail: gkbakshi73uro@hotmail.com

ABSTRACT

Extrarenal primary renal cell carcinoma is an extremely rare entity with limited literature. Characteristic findings of renal cell carcinoma (RCC) at the metastatic site in the absence of a discrete radiological renal lesion can perplex both the clinician and pathologist. We report a case of metastatic primary extrarenal papillary RCC, who presented as a neck mass clinically and radiologically a paraaortic mass with normal bilateral native kidneys. The final diagnosis was aided by histopathological features, further confirmed by targeted immunohistochemical markers.

INTRODUCTION

Renal cell carcinoma (RCC), which includes 2%–3% of all adult solid tumors, are metastatic at the presentation in 30%% - 40% of cases, with lungs being the most common.[1] There are various reports of RCC metastasis in the head-and-neck regions such as sino nasal, orbit, lacrimal gland, tongue, tonsils, parotid, and thyroid.[1] This is attributed to the expression of inducible oncogenes that bypass the primary metastatic site to reach unusual secondary sites.[1] We report a case of primary extrarenal papillary carcinoma with multiple paraesophageal, mediastinal and supraclavicular nodal metastasis presenting as a left sided neck mass.

CASE REPORT

A 34-year-old male without comorbidities, addictions and no significant family history presentedwith a new-onset left-sided neck swelling. He had a good performance status and was pale. Abdominal examination was normal. OOn examination of the neck, a 4 cm × 3 cm nontender, firm, andimmobile, conglomerated lymph nodal mass was palpable in the left supraclavicular region [Figure 1a]. Oral cavity and thyroid were normal. Scrotal examination revealed normal bilateral testes. There were no other sites of lymphadenopathy.

On PET-CT, an FDG avid, hypo enhancing paraaortic mass 64mm x 66mm [SUVmax] : 11.43) with areas of necrosis, encasing and displacing the left renal vein and artery anteriorly [Figure 1b] and abutting the posterior surface of thethe body of pancreas was noted. FDG avid mediastinal, paraesophagal and prevascular nodes [Figure 1c] and a left supraclavicular nodal mass measuring 4.5 cm x 4.8 cm were noted. Buckling and denting of the left renal cortex by the mass and presence of a discrete fat plane between the renal hilum, upper ureter and the mass confirmed the extrarenal origin [Figure 1d-f]. Left adrenal was separate and normal. The mass had no ureter or blood supply, which ruled out the possibility of a supernumerary kidney. The ultrasonogram of the neck showed a normal thyroid and left supraclavicular nodal mass.

CT-guided biopsy of the paraaortic mass done elsewhere and reviewed by a uropathologist and showed papillae lined by

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pseudostratified layers of cells with abundant eosinophilic cytoplasm and higher nucleolar grade suggestive of papillary carcinoma [Figure 2a]. On immunohistochemistry (IHC), the tumor was strongly and diffusely positive for paired box 8 (PAX8) [Figure 2b], exhibited staining for alpha-methyl CoA racemase (AMACR) [Figure 2c], and was negative for transcription factor for immunoglobulin heavy-chain enhancer 3 (TFE 3) and thyroid transcription factor 1.

Based on this ancillary IHC, the diagnosis was confirmed to be extrarenal papillary RCC type II. In view of the metastatic stage, subtype, and Intermediate risk IMDC score (International Metastatic Renal Cell Carcinoma Database Consortium) the patient was counseled about palliative intent of treatment in the multidisciplinary tumor board and started on tyrosine kinase inhibitors due to non-affordability for immunotherapy. The patient was unable to continue management in our institute and hence the response could not be assessed.

DISCUSSION

Ectopic solid tumors have been reported previously. These include functional tumors such as adrenocorticotropic hormone secreting tumors and non-functional tumors. Primary RCC has been described in patients with ectopic and supernumerary kidneys, but it is rarely detected in an extrarenal tissue with normal bilateral kidneys.

Previously, four similar cases with variable histologies have been reported in the literature. Terada[2] reported one such case which was an incidental radiological finding in a follow-up case of carcinoma rectum, while Hasan *et al*.[3] reported a post-operative finding following the resection of a presumed adrenal mass. Al-Maghrabi *et al*[4] and Youjian Li *et al*[5] reported similar cases . While all these cases were nonmetastatic and resectable at presentation, our case is the first extrarenal primary RCC with synchronous metastases not amenable for upfront surgery [Table 1].

Extrarenal RCC originates from an abnormally located renal tissue without a separate collecting system and in the presence of normal native kidneys. During the fetal kidney development, the metanephros persists as metanephric blastema by the 7th week of gestation, evolving into the normal postnatal kidneys. During this differentiation, some embryonic mesonephric remnants can remain postnatally[6] and its subsequent tumor formation can present diversely.

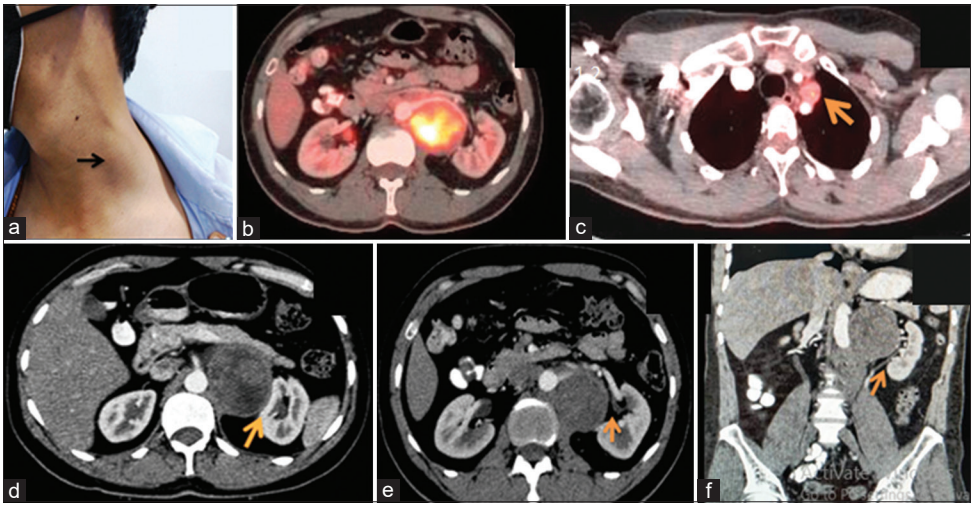

Figure 1: Clinical photograph and Imaging. (a) Left supraclavicular Nodal mass; (b and c) PET CECT – FDG avid (SUVmax : 11.43) extra renal mass and the metastatic mediastinal node; (d and e) CECT Axial section - buckling and denting of the Left renal cortex by the mass; (f) CECT Coronal section - the presence of a discrete fat plane between the renal hilum upper, ureter and the mass

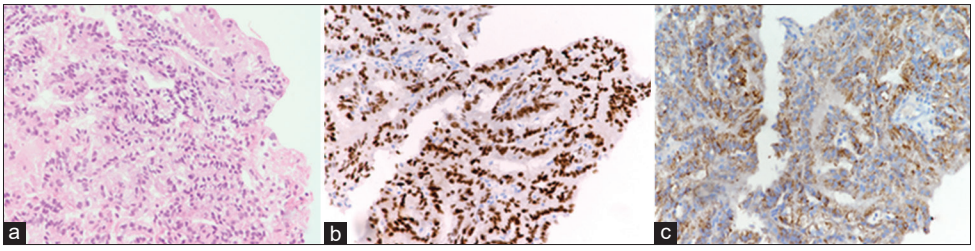

Figure 2: Histopathology and immunohistochemistry photograph (a) histopathology photograph paraaortic mass biopsy on H and E (×200), (b) immunohistochemistry with paired box 8 positivity (×400), (c) immunohistochemistry with alpha-methyl CoA racemase positivity (×400)

Table 1: List of cases reported in literature

Presentation	Stage	Management	Histopathology	Author
Incidental radiological finding	NM	Surgery with curative intent	cRCC	Terada *et al*., 2012[2]
Abdominal mass	NM	Surgery with curative intent	cRCC	Hasan *et al*., 2015[3]
Abdominal mass	NM	Surgery with curative intent	Xp11 Translocation associated RCC	Al-Maghrabi JA *et al*., 2017[4]
Abdominal pain	NM	Surgery with curative intent	Type II pRCC	Youjian Li *et al*., 2019[5]
Neck mass	Metastatic	CT guided Biopsy and TKIs with palliative intent	Type II pRCC	Our case

TKIs=Tyrosine Kinase Inhibitors, NM=NonMetastatic, RCC=Renal cell carcinoma, cRCC=Clear cell RCC, pRCC=Papillary RCC

Papillary RCC has a propensity to involve nodes (13%) compared to clear cell RCC (8.6%)[6] and also enhance to a lesser extent which is an important differentiating feature on CECT. The American College of Radiology guidelines suggest an FDG PET CT if the mediastinal node was above 15 mm in short axis diameter without an explicable disease. Indeterminate lymphadenopathy in the mediastinum, retroperitoneum, and supraclavicular should be differentiated using radiological and histopathological features. In such cases, a gun needle biopsy with ancillary IHC would be helpful toward confirming a diagnosis.

PAX8 is expressed in the embryogenesis of thyroid, Müllerian, renal/upper urinary tracts[7] and the frequency of expression in the tumors arising from these tissues is up to 91%. In case of an unknown primary tumor, a second specific marker AMACR is used and has a frequency of expression of 84% in papillary RCC.[8] PAX2, CD10, cytokeratin, and vimentin are also used. A proper staging workup must be made as for a renal mass. Even though there is no standard staging classification for such tumors the same line of management and prognostication of a primary papillary RCC is recommended.

CONCLUSION

To conclude, we report a rare case of metastatic primary extrarenal papillary carcinoma. It is a diagnostic challenge due to the lack of pathognomonic features and unusual presentation. Hence, awareness of the possibility of an extrarenal malignancy and the reliance on imaging with IHC is emphasized.

Declaration of patient consent

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

REFERENCES

1. Sountoulides P, *et al*. Atypical presentations and rare metastatic sites of renal cell carcinoma: A review of case reports. Journal of Medical Case Reports 2011;5:429.
2. T Terada, *et al* , Extra-renal clear cell renal cell carcinoma probably arising from mesodermal embryonic remnants - Pathology international, 2012.
3. R Hasan, S Kumar, V Monappa Primary extra-renal clear cell renal cell carcinoma masquerading as an adrenal mass: A diagnostic challenge Urology annals, 2015;7(4):513-5.
4. Al-Maghrabi JA *et al*. Uncommon localization of extrarenal Xp11.2 translocation-associated renal cell carcinoma (RCC): Case report. Appl Immunohistochem Mol Morphol 2017; e33–5.
5. Youjian Li , Xuefeng Qiu , Weijian Li , Yang Yang , Rong Yang , Xiaozhi Zhao. Primary Extrarenal Type 2 Papillary Renal Cell Carcinoma: A Case Report Journal of Urology 2018: 10.039.Volume 123, E1 – E3.
6. Raghunandan Vikram, Chaan S, Pheroze Tamboli, Nizar Tannir, Eric Jonasch, Surena Matin *et al*,. Papillary Renal Cell Carcinoma: Radiologic-Pathologic Correlation and Spectrum of Disease, Radio Graphics (RSNA) May 2009; 29:741–757.
7. Laury AR, Perets R, Piao H, Krane JF, Barletta JA, French C, *et al*.. Comprehensive analysis of PAX8 expression in human epithelial tumours. American Journal of Surgical Pathology. 2011 Jun; 35(6):816-26.
8. Eichelberg C, *et al*.. Prognostic value of alpha-methyl CoA racemase (AMACR) expression in renal cell carcinoma. World Journal of Urology. 2013 Aug; 31(4):847-53.

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