Case Report

Extranodal Rosai–Dorfman disease: A rare mimicker of metastatic renal cell carcinoma

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ABSTRACT

A 29-year-old female presented with the history of pain in the abdomen and a palpable lump in the right hypochondrium, lumbar, and the umbilical regions for the past 2 months. On evaluation with contrast-enhanced computed tomography (CECT) and positron emission tomography CT, she was found to have a heterogeneously enhancing mass entirely replacing the mid and the lower pole of the right kidney, the pelvis, and the upper ureter with loss of fat planes with the inferior vena cava, psoas muscle, and the hepatic flexure, along with pericardial deposits and soft tissue lesions at multiple paravertebral regions and the right thigh. Owing to a high suspicion of metastatic renal cell carcinoma (RCC), a right cytoreductive nephrectomy was performed. Histopathology revealed extranodal Rosai–Dorfman disease, which was mimicking a metastatic RCC on imaging. The patient was started on oral steroids to control the distant lesions and to prevent progression of the disease and is doing well at follow up.

INTRODUCTION

Renal cell carcinoma (RCC) accounts for approximately 2% of the global cancer burden and 30% of the patients present with a metastatic disease.[1] The diagnosis can only be made on histopathology and patients with typical imaging findings of metastatic RCC may rarely have a completely different disease altogether. Herein, we report a patient with imaging findings classically suggestive of metastatic RCC which turned out to be extranodal Rosai–Dorfman disease (RDD).

CASE REPORT

A 29-year-old female presented to the outpatient department with the complaints of intermittent, dull aching, non-radiating pain in the right flank associated with vomiting for the past 2 months. There was no history of hematuria, dysuria or fever, the appetite was normal and she did not complain of weight loss. The general physical examination was unremarkable.

On the abdominal examination, a large 10 cm × 8 cm firm ballotable lump was palpable in the right hypochondrium, extending up to the right lumbar and the umbilical region. She was further evaluated and the ultrasonography of the abdomen revealed a 17 cm × 9 cm heterogeneous mass lesion with internal vascularity involving the lower half of the right kidney. The contrast-enhanced computed tomography (CECT) scan of the abdomen and thorax revealed a heterogeneously enhancing soft tissue density mass lesion of size 12.6 cm × 13.2 cm × 15.4 cm arising from the right kidney, pushing and compressing the renal cortex, with extension into the right retroperitoneal space displacing the ascending and the transverse colon anteriorly and the small bowel loops and the stomach toward the left side [Figure 1], with suspicious soft-tissue deposits along the left thoracic paravertebral space and in the pericardium along the right atrium. A provisional diagnosis of a metastatic right-sided RCC was made and the patient underwent a FDG positron emission tomography scan which corroborated the findings of the CECT scan with a

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SUV_{max} of 2.0 in the primary lesion and 1.9 in the metastatic lesions. It also revealed an additional heterogeneously FDG avid subcutaneous soft tissue thickening 5mm x 5mm in size along the medial aspect of the right thigh. In view of these findings, she underwent a right cytoreductive nephrectomy through a thoraco-abdominal approach. The cut section of the specimen revealed a large heterogeneous tan brown tumor of 15 cm × 15 cm size replacing the entire middle and the lower pole with a small area of normal renal cortex at the upper pole. The tumor was limited to the Gerota's fascia and the renal vein was free [Figure 2]. The histopathological examination revealed large collections of histiocytes with an enlarged round to oval nuclei and abundant pale staining cytoplasm. These histiocytes were exhibiting emperipolesis [Figure 3]. Abundant infiltration of adipocytes, plasma cells, and lymphocytes was also seen along with histiocytes. The renal parenchyma showed few collections of histiocytes along with occasional lymphoid follicles with germinal center formation. The cells were positive for S-100, CD 31, CD 138, CD 68, and negative for Pan CK, SMA, and Alk. In view of the morphological and the immunohistochemical features, a diagnosis of extranodal RDD was made. The case was discussed in a multi-disciplinary tumor board and the patient was started on steroids after consultation with the immunologist. Currently, the patient is doing well at 3 months' follow-up.

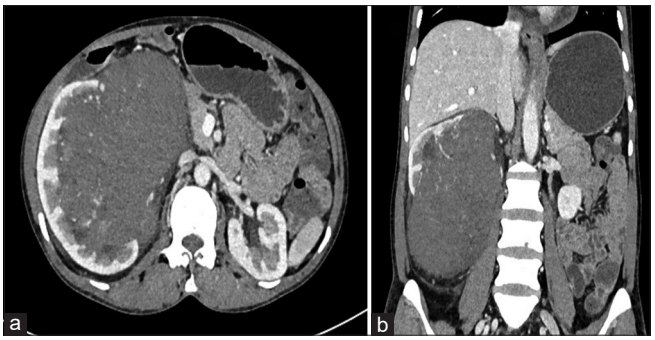

Figure 1: Contrast-enhanced CT scan of the abdomen, axial (a) and coronal (b) sections, showing the large enhancing right renal mass with internal vascularity, replacing almost the entire renal parenchyma

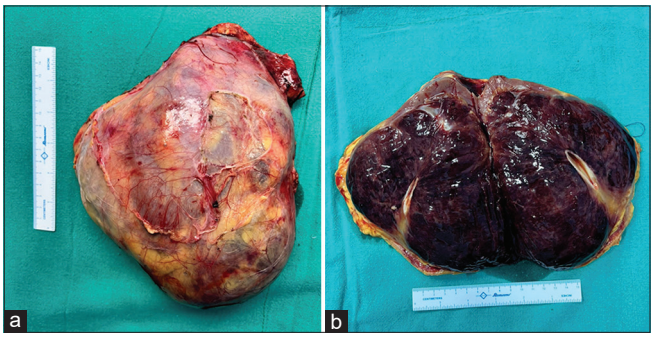

Figure 2: Right radical nephrectomy specimen (a) Cut Section (b) showing a large tan brown tumor with a thin rim of remaining parenchyma at the upper pole

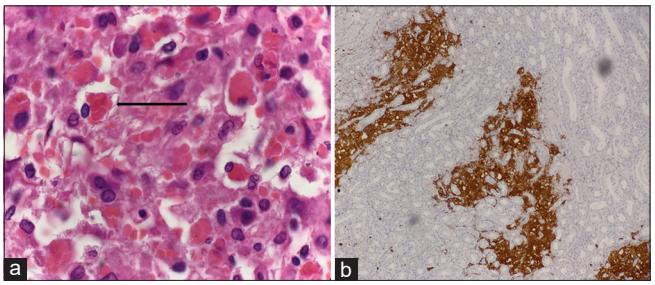

Figure 3: Microscopic examination (a) H and E, ×40 showing large collections of histiocytes with an enlarged round to oval nuclei and abundant pale staining cytoplasm. These histiocytes are exhibiting emperipolesis (arrow). (b) Immunohistochemistry staining with S-100 at ×10, showing positive tumor cells in a background of normal renal tubules

DISCUSSION

Our patient presented to the OPD with a large palpable right renal lump and the imaging features suggestive of a heterogeneously enhancing renal mass with metastases. Metastatic RCC was invariably kept at the top of the list of differential diagnoses with others being round cell tumor/lymphoma or renal sarcoma. Tuberculosis has also been reported to mimic metastatic RCC.[2] The classical sites of metastasis in RCC are the lungs, bones, lymph nodes, and the liver. However, our patient had metastases in the pericardium, paravertebral space and the skin. Although rare, metastasis to these locations have been previously reported in patients with RCC.[3] Considering the fact that the patient had a large palpable symptomatic mass, replacing almost the entire renal parenchyma, probably rendering it nonfunctional, a preoperative renal mass biopsy was not perfromed and she was planned for, and underwent a cytoreductive nephrectomy, based on the International Metastatic RCC Database Consortium risk criteria.

Histopathology revealed a diagnosis of RDD and the case was reviewed retrospectively. Renal involvement in RDD is rare, as is the presentation as a metastatic renal mass and hence RDD was not one of the initial differential diagnoses. Similar to earlier reports, the patient was treated as a case of malignant renal mass and underwent nephrectomy without suspicion of histiocytic tumors.[4-6] The pattern of metastases in our patient was atypical, which was a marker that we could have been dealing with an altogether different histopathology rather than a straightforward metastatic RCC.

As per the revised classification of histiocytoses, RDD is classified in the "R" group, including the familial, classical, extranodal, neoplasia-associated, and the immune disease-associated RDD.[7] It was initially reported as sinus histiocytosis with massive lymphadenopathy,[8] and is a rare, benign histiocytic proliferation disorder, characterized classically by painless cervical lymphadenopathy which may be associated with preceding fever, malaise, throat pain, night sweats, or weight loss.[9] It usually presents in the 2nd or the 3rd decade of life and up to 43% of the patients are reported to have an extranodal involvement, most commonly

in the skin, the nasal cavity and the paranasal sinuses, the eyes, adnexa and the bones.[8] Histopathology classically reveals large histiocytes with ample pale or "watery-clear" cytoplasm with a large hypochromatic nucleus and a prominent nucleolus and usually exhibit emperipolesis but are not pathognomonic. The immunophenotype of histiocytes in RDD is characterized by cytoplasmic and nuclear S100 and fascin positivity, with CD68 and variable CD163 and CD14 positivity. The cells are CD1a and CD207 negative in contrast to Langerhans cell histiocytosis [8] Our patient exhibited these features which led to the diagnosis of RDD. Renal involvement in RDD is rare and portends a poor prognosis.[9] These patients may present with hematuria, flank pain, abdominal fullness, or nephrotic syndrome.[8]

Differential diagnoses of renal RDD include Erdheim–Chester disease, renal lymphoma, IGG-4-related disease, tuberculosis or metastatic tumors. Xanthogranulomatous pyelonephritis has been reported to mimic RCC and upper tract transitional cell carcinoma and should also be included in the list of differentials. In patients presenting with metastatic renal tumors, the presence of metastasis at atypical sites should prompt an investigation into the other possible differential diagnoses. The diagnosis of RDD is confirmed on histopathology which reveals large histiocytes with pale staining cytoplasm, exhibiting emperipolesis and S-100 positivity.[8] Our patient exhibited all these features and was confirmed to have RDD.

Multiple treatment modalities have been tried in patients with RDD including, but not limited to steroids, chemotherapy and radiotherapy, with mixed results[10] and the standard of care remains to be defined. We started the patient on oral steroids after discussion in the tumor board and currently, the patient is doing well, with no signs of worsening or disease progression.

CONCLUSION

In patients with large renal masses with metastases at the atypical sites, alternate diagnoses should be suspected and a renal mass biopsy should be performed as the findings of a benign pathology can alter the course of management. However, in certain situations like ours, where the patient has a large symptomatic renal mass, probably leading to a nonfunctional kidney, a renal mass biopsy may be avoided and a nephrectomy can be performed with a cytoreductive intent.

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. Tran J, Ornstein MC. Clinical review on the management of metastatic renal cell carcinoma. JCO Oncol Pract 2022;18:187-96.
2. Ko Y, Mo EK, Park YB, Kang MR, Bae JS, Kim Y. Miliary Tuberculosis Mimicking Brain Metastasis from Renal Cell Carcinoma. J Neurocrit Care 2018;11:47-53.
3. Sountoulides P, Metaxa L, Cindolo L. Atypical presentations and rare metastatic sites of renal cell carcinoma: A review of case reports. J Med Case Rep 2011;5:429.
4. Tanaka H, Mukai S, Kamoto T, Kataoka H. Extranodal Rosai-Dorfman disease of the kidney: A case report. Hum Pathol Case Rep 2019;17:200306.
5. El Majdoub A, El Houari A, Chbani L, El Fatemi H, Khallouk A, Farih MH. Isolated localization of Rosai Dorfman disease as renal mass: A case report and review of literature. Pan Afr Med J 2016;24:64.
6. Abdollahi A, Ardalan FA, Ayati M. Extranodal Rosai-Dorfman disease of the kidney. Ann Saudi Med 2009;29:55-7.
7. Emile JF, Abla O, Fraitag S, Horne A, Haroche J, Donadieu J, *et al.* Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages. Blood 2016;127:2672-81.
8. Abla O, Jacobsen E, Picarsic J, Krenova Z, Jaffe R, Emile JF, *et al.* Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. Blood 2018;131:2877-90.
9. Gaitonde S. Multifocal, extranodal sinus histiocytosis with massive lymphadenopathy: An overview. Arch Pathol Lab Med 2007;131:1117-21.
10. Goyal G, Ravindran A, Young JR, Shah MV, Bennani NN, Patnaik MM, *et al*. Clinicopathological features, treatment approaches, and outcomes in Rosai-Dorfman disease. Haematologica 2020;105:348-57.

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