Case Report

Rosai Dorfman Disease of Kidney: Report of a Rare Case

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Abstract

Rosai Dorfman Disease (RDD) is a type of histiocytoses usually affecting the lymph nodes. Extranodal involvement by RDD can occur, although, infrequently. However, extranodal RDD without a prior history of the nodal variant of the disease is rare. Present case report highlights this rare occurrence. A 75 years old female presented with left renal mass with no significant previous medical or surgical history. PET-CT scan revealed two solid lobulated mass in the left kidney, suggesting Renal Cell Carcinoma. Radical nephrectomy was performed, which on histopathological examination showed features of RDD and was confirmed subsequently by immunohistochemical and special stains.

Keywords:

Rosai Dorfman Disease, Kidney, Histiocytosis, Renal cell carcinoma

Introduction

Rosai Dorfman Disease (RDD) also known as Sinus histiocytosis with massive lymphadenopathy (SHML) is one of the five types of histiocytoses classified as R Group histiocytoses. First described in 1969, this disease, in its classical sporadic form presents in children and young adults with bilateral painless lymphadenopathy in the neck along with fever, night sweats, fatigue, and weight loss.[1]

Lymph nodes are the most commonly involved site; however, any organ may be affected. Extranodal involvement by sporadic RDD has been documented in 43% of cases; the most frequent sites being skin, nasal cavity, bone, soft tissue, and retro-orbital tissue. Thus, the term RDD has been adopted in place of SHML. [2,3]

Case Report

A 75-year-old female presented with right flank fullness and pain since 2 months. The patient had no significant previous medical or surgical history. On physical examination, tenderness was noted in the right lumbar fossa. There was no peripheral lymphadenopathy, no history of fever or significant weight loss. Hematological examination revealed microcytic hypochromic anemia (haemoglobin= 7.0 g/dl) and raised Erythrocyte sedimentation rate (ESR) (46 mm/hour). Prothrombin time and international normalized ratio (INR) were slightly increased to 16.9 seconds and 1.3 respectively. Routine and microscopic urine examination revealed traces of blood (0-1/high power field) with no other abnormal finding. Renal function test, liver function test and blood sugar levels were within normal limits.

On radiological examination, Positron emission tomography and computed tomography (PET-CT) scan revealed two solid lobulated mass of sizes 5.1×3.7×4.6 cm and 4.0×3.3×3.8 cm in mid and lower pole of left kidney, infiltrating renal pelvis infundibulum and upper pole calyx, suggesting Renal Cell Carcinoma (Figure.1). Also, non-fluorodeoxyglucose (FDG) avid multiple sub centimetric, aortocaval, preaortic, paraaortic and mesenteric lymph nodes (reactive/ inflammatory) were identified. Radical nephrectomy was done with the provisional diagnosis of Renal Cell Carcinoma.

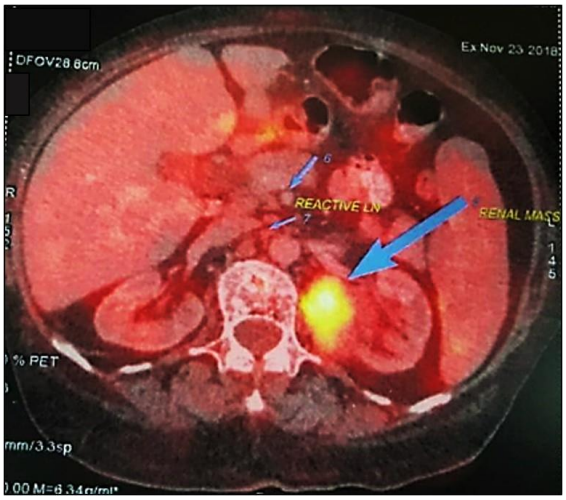

Figure 1 PET-CT image showing solid lobulated mass in mid & lower pole of left kidney, infiltrating renal pelvis infundibulum and upper pole calyx

Grossly, the left kidney measured 11×6×5 cm. On cut section, a multinodular pale yellow lesion involving predominantly the lower and middle pole, renal sinus and pelvicalyceal system measuring 5.2×3.1×3.6 cm was identified (Figure 2). The lesion merged imperceptibly with the renal parenchyma and was seen to infiltrate the renal capsule. Microscopic examination from the lesion showed sheets of histiocytes in a background of chronic inflammatory infiltrate comprising predominantly of plasma cells and lymphocytes. These histiocytes had abundant granular eosinophilic cytoplasm, vesicular nuclei and smooth nuclear contours. Extensive emperipolesis was noted (Figure 3 a, b).

Immunohistochemical staining showed expression of S100 and CD68 in these histiocytes whereas CD1a was negative ((Figure 3c). Special stains performed included Zeihl-Neelsen stain, PAS, Von kossa and Perls Prussian blue stains, which were all negative. Based on the histomorphological and immunohistochemical findings, a final diagnosis of extranodal RDD involving

kidney was given.

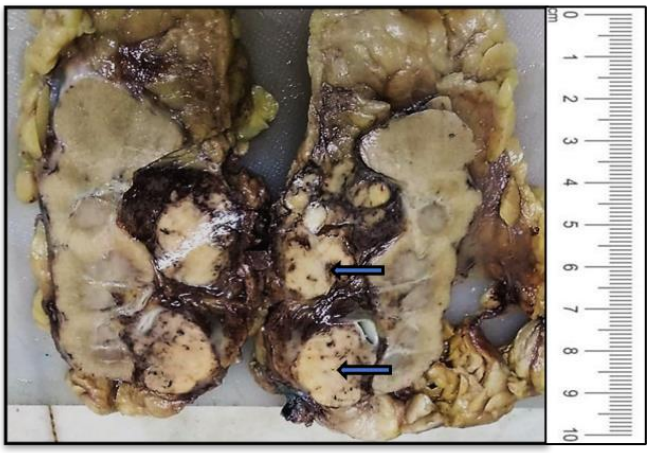

Figure 2 Gross appearance of the nephrectomy specimen showing yellowish-brown lesion in lower & mid pole of kidney (arrows) infiltrating into the capsule

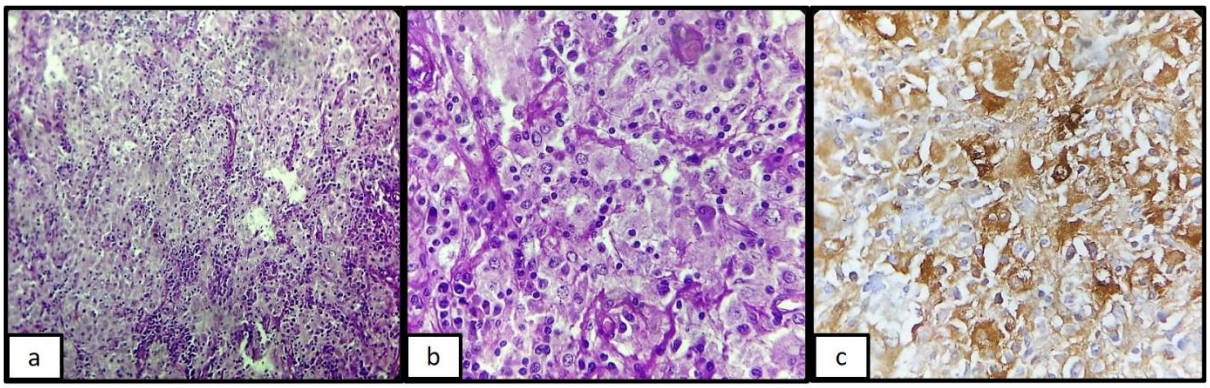

Figure 3 (a) Sheets of histiocytic cells along with numerous lymphocytes and plasma cells (Hematoxylin & Eosin stain 200X); (b) Higher magnification shows extensive emperipolesis and dispersed lymphocytes and plasma cells (Hematoxylin & Eosin stain 600X); (c) Immunohistochemical staining with S-100 highlighting strongly stained histiocytes (600X)

Discussion

RDD is a rare benign histiocytic disorder. The etiology is unclear, although autoimmune diseases, chronic inflammatory cause and viruses such as human herpesvirus 6 or Epstein-Barr virus have been postulated as potential sources.[4-6] Recent studies have demonstrated the role of the RAS/RAF/ERK (MAPK) pathway in RDD, and 33% of cases had mutually exclusive KRAS and MAP2KA mutations.[7]

RDD exhibits a slight male predominance (58%) and a general predilection for individuals of African descent.[8] Laboratory findings include anemia, leukocytosis and serum polyclonal hypergammaglobulinemia, hyperferritinemia.

In RDD, lymph nodes are the most commonly involved site; however, any organ may be affected. Extranodal involvement by sporadic RDD has been documented in 43% of cases; the most frequent sites being skin, nasal cavity, bone, soft tissue, and retro-orbital tissue.[9]

Kidney involvement in RDD is very uncommon, and therefore RDD is not frequently considered in the differential diagnosis of an infiltrative renal mass. Renal RDD occurs in sixth to seventh decade age group. Coexistent lymphadenopathy, especially in retroperitoneal sites such as the para-aorta, iliac and inguinal LN has been noted in 78% of patients. The simultaneous involvement of more than one site in the same individual is observed in upto 44.7% of cases.[10]

The histological findings in renal RD are characterized by dense infiltrate of histiocytes with scattered lymphocytes, plasma cells, and neutrophils. These histiocytes have large vesicular nuclei, inconspicuous nucleoli, and abundant pale eosinophilic cytoplasm; showing emperipolesis, which is the pathognomic feature of RDD. Immunohistochemistry shows positivity for S-100 and CD68 and negativity for CD1a. [9]

Differential diagnosis of histiocytic infiltrative lesion in kidney includes xanthogranulomatous pyelonephritis (XPG), Tuberculosis (TB) Kidney, Malakoplakia, Langerhans cell histiocytosis (LCH), Erdheim Chester Disease, Histiocytic sarcoma. [9,11] Absence of emperipolesis and IHC profiles and special stains help to reach to a correct diagnosis.

In XPG, fibrosis and chronic granulomatous inflammatory infiltrate with lipid-laden macrophages are found, but emperipolesis and S100 positivity is absent. In TB Kidney, epithelioid cell granulomas with mononuclear inflammatory infiltrate are seen which may be accompanied with AFB positivity and caseous necrosis. These features are sufficient to distinguish these conditions from RDD.

In Malakoplakia, soft, yellow–tan tissue may replace large areas of renal parenchyma along with prominent fibrosis and may be confused with RDD as the inflammatory infiltrate is predominantly histiocytic accompanied with few lymphocytes and plasma cells. However, the characteristic Michaelis–Gutmann bodies which are positive for PAS, Von kossa and Perls Prussian blue, help in differentiating malakoplakia from RDD.

Langerhans cell histiocytosis (LCH) also can be considered as a differential for RDD. However, the coffee bean appearance of the cells and positivity for both S100 and CD1a in LCH, is useful in its definitive diagnosis. Similarly, Erdheim Chester Disease (ECD) is one of the differentials for RDD morphologically as in ECD, clonal systemic proliferation of histiocytes occurs, usually having a foamy (xanthomatous) component. But the presence of Touton giant cells with accompanying osteosclerosis is a differentiating feature helpful in reaching diagnosis.

Other possible differential diagnoses include storage disease, metastatic tumor such as malignant melanoma of clear cell type or even renal cell carcinoma.[9]

RDD generally has a favorable prognosis, but involvement of extranodal systems worsens the prognosis with a higher tendency for a chronic relapsing course and higher mortality rate when associated with involvement of unusual sites such as kidney.[11]

On extensive literature search, there have been less than 15 case reports of extranodal RDD presenting as a renal mass in an elderly patient without a prior history of the nodal variant of the disease.

Conclusion

RDD of the kidney, although rare, should be considered in the differential diagnosis of renal mass appearing as renal cell carcinoma in computed tomography scan to avoid unnecessary radical nephrectomy.

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Competing Interests: none

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