

Extranodal Rosai-Dorfman disease of the kidney

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CASE SUMMARY

A 28-year-old female with no significant medical history presented with abdominal pain, nausea, and vomiting, along with left-sided abdominal mass of approximately 6 months' duration with progressive enlargement. The patient denied any loss of appetite, weight loss, cough, chest pain, shortness of breath, or urinary or bowel symptoms. Physical exam revealed a firm, palpable mass in the left hemi-abdomen. No direct or rebound tenderness was appreciated. Laboratory evaluation showed mild iron deficiency anemia.

IMAGING FINDINGS

A CT scan of the abdomen demonstrated a $9.3 \times 8.2 \times 17.5$ cm heterogeneously enhancing left renal mass, with significant mass effect and displacement of adjacent structures. The left renal vein and inferior vena cava were patent without evidence of intraluminal filling defect or tumor thrombus. Left para-aortic lymphadenopathy was also noted, with the largest lymph node measuring 2.2×1.8 cm (Figure 1). Retroperitoneal ultrasound showed a large heterogeneous solid mass with increased color Doppler flow. There was calyceal dilation noted with a rind of renal cortical tissue along the superior aspect of the mass (Figure 2). A CT-guided biopsy of the renal mass lesion was subsequently performed by interventional radiology.

Pathology sections from the left kidney demonstrated a lesion composed predominantly of histiocytes with admixed plasma cells and lymphocytes and focal evidence of emperipolesis. Immuno-stains showed histiocytes

to be CD68 positive and negative for CD1A. S-100 showed diffuse positivity in the histiocytes. Cytokeratin was negative and PAX-8 highlighted background lymphocytes. The presence of emperipolesis and S-100 immunostaining are most consistent with extranodal Rosai-Dorfman disease (Figure 3).

Patient was referred to surgery for left radical nephrectomy via a sub-costal incision. Intra-operative findings demonstrated a bulky renal mass, which was removed en bloc.

DIAGNOSIS

Extranodal Rosai-Dorfman disease

DISCUSSION

Rosai-Dorfman disease (RDD), also referred to as sinus histiocytosis with massive lymphadenopathy (SHML), is a rare, benign proliferative histiocytic disorder first described in 1969 by Rosai and Dorfman.^{1,2} The most common presentation of Rosai-Dorfman disease is painless lymphadenopathy; most commonly bilateral cervical with constitutional symptoms. In about 43% of patients the disease may also arise in extranodal sites and almost any organ can be involved.³

The imaging findings of RDD are very nonspecific, with a wide differential diagnosis which includes benign and malignant neoplasm, infection and other mass lesion etiologies. As such, diagnosis depends on tissue sampling and pathology demonstrating diffuse histiocytosis with presence of emperipolesis and S-100 immunostaining.⁴

Genitourinary, specifically renal involvement has been rarely described.

A case of an adult female with a retroperitoneal pelvic mass causing urinary obstruction was described by Leiva, et al.⁵

Manifestations of extranodal RDD have been treated with surgical resection, medical/chemotherapy and radiation therapy; however surgical therapy has been the most successful and definitive treatment.^{5,6} The most commonly reported sites of extranodal involvement are the skin and soft tissue; this usually presents as well-defined papules to palpable masses, with or without satellite lesions.⁷

Our patient presented with a slowly growing left sided abdominal mass over a period of 6 months. Emergency presentation was precipitated by sudden pain over the left side of the abdomen. During initial clinical evaluation, there were no systemic constitutional symptoms described or lymphadenopathy other than a palpable left sided abdominal mass. A CT scan of the abdomen did demonstrate a large left renal mass and multiple enlarged para-aortic lymph nodes measuring up to 1.8 cm in short axis.

Our case demonstrated classic histological features suggestive of RDD, which included emperipolesis (presence of an intact cell within the cytoplasm of another cell) and diffuse positivity for S-100 immuno-stains. Surgical resection was chosen due to the size of the mass and associated symptomatic mass effect.

CONCLUSION

Extranodal Rosai-Dorfman syndrome accounts for almost half of all clinical presentations; however, renal involvement has been very rarely reported. We

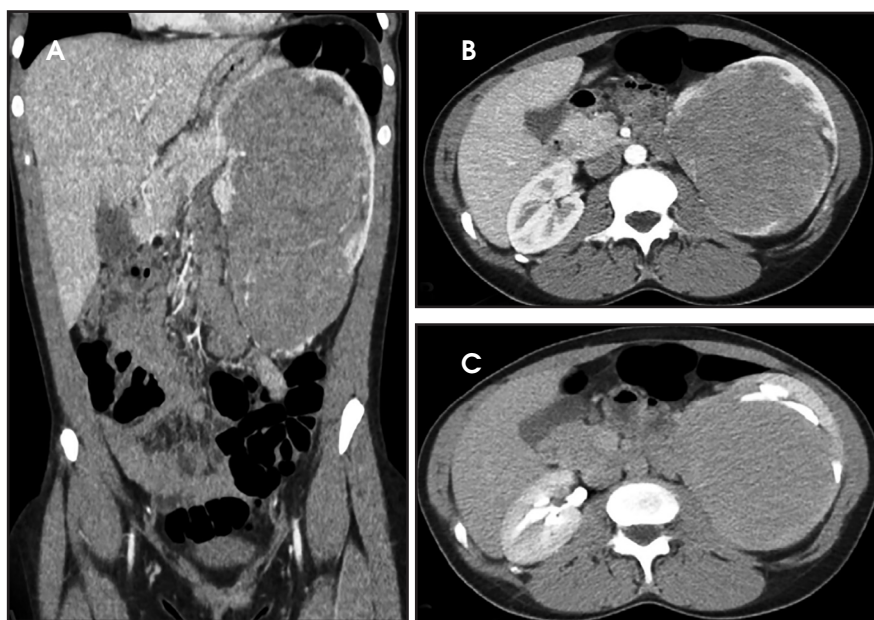


FIGURE 1. Coronal (A) and axial (B, C) contrast-enhanced CT imaging of the abdomen demonstrate a large heterogeneous mass arising from the left kidney showing mild enhancement splaying and displacing the surrounding renal cortex. There is mass effect noted with anterolateral displacement of the renal collecting system as seen on the excretory phase of the CT scan (C). Multiple enlarged para-aortic lymph nodes are also seen.

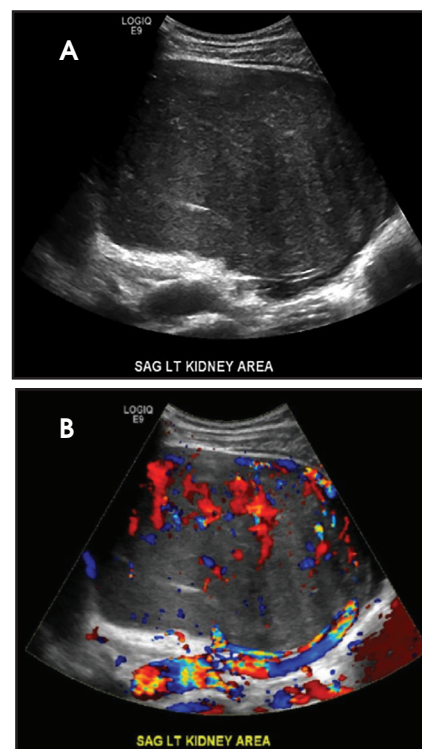


FIGURE 2. The left kidney appears replaced by an 18.9 × 10.1 × 12.2 cm solid mass with internal color flow. There appears to be a rim of renal cortical tissue in the upper pole with a moderately dilated calyx.

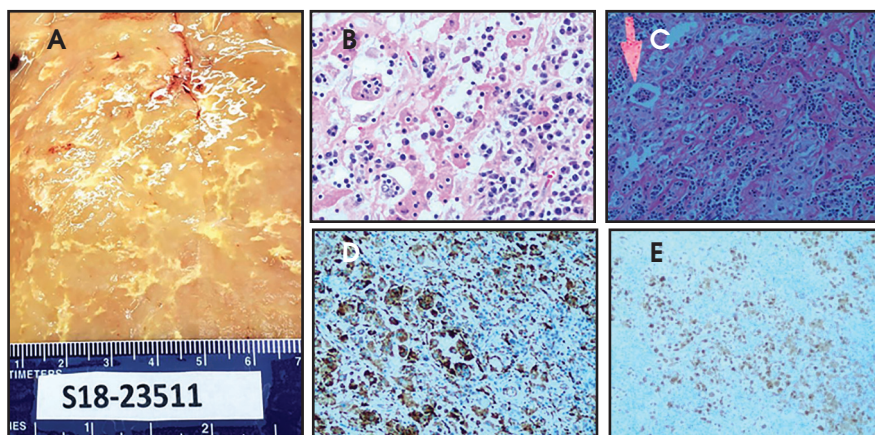


FIGURE 3. (A) A gross post-surgical specimen demonstrating a yellow hue glistening texture; (B, C) 10X H&E microscopic picture demonstrating histiocytes with admixed plasma cells and lymphocytes and focal evidence of emperipolesis (red arrow); (D) CD68 stain 10X microscopic picture; (E) S100 stain 4X microscopic picture shows diffuse positivity in the histiocytes, highlighting rare emperipolesis.

present a case of extranodal RDD as a renal mass in a young female patient, with imaging findings and pathologic-diagnosis correlation.

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