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Rosai-Dorfman-Destombes disease with renal involvement and secondary glomerulopathy: Report of an exceptional case

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Introduction

Rosai-Dorfman-Destombes disease (RDD) or sinus histiocytosis with massive lymphadenopathy (SHML), is a rare disorder, first described by Destombes in 1965 and then Rosai and Dorfman reported the first clinical series in 1969. The etiology of RDD is unknown, and the treatment is not standardized. There is no data about the true number of cases that exist worldwide. This disease occurs mainly in children and young adults, however, it may occur at any age. It is more common in males and individuals of African descent.

RDD, presents clinically as painful bilateral massive cervical lymphadenopathies, with fever, night sweats and weight loss. Mediastinum, inguinal region and retroperitoneum lymph nodes may also be involved. The course of the disease is unpredictable, usually characterized by episodes of exacerbation and remission. It can last for many years and is usually self-limited, with good outcomes. Extranodal compromise is unusual but can affect almost any organ. Involvement of the kidney has been stated as a poor prognostic predictor.[1]

We present an unusual case of RDD in a 64 year old woman with a literature review of renal RDD.

Case report

A 64-year-old woman with hypertension and Sjogren's syndrome, was evaluated in May 2014 because of chronic diffuse abdominal pain, without any other complaints. An abdominal computerized tomography (CT) showed a left renal pelvic lesion suggestive of transitional cell carcinoma. To further characterize the lesion, an abdominal magnetic resonance (MRI) was performed, which showed a mass of 5.0× 2.4 cm in the left renal pelvis, associated with moderate hydronephrosis (Fig. 1). It also informed multiple bilateral perirrenal adenopathies, up to 13 mm in diameter. Main laboratory findings were serum creatinine of 1.06 mg/dL, (eGFR 52 ml/min/1,73 m^2 by MDRD) and ESR of 55 mm/hr. A laparoscopic left nephroureterectomy was performed. The biopsy informed a polymorphic inflammatory infiltrate, with lymphocytes, plasmocytes, eosinophilic and histiocytic large cells with intracytoplasmic lymphocytes (emperipolesis) (Fig. 2), replacing the renal cortex (Fig. 3).

In the immunohistochemistry, the histiocytes were strongly positive for S100 protein, CD163 and CD 68. The lesion was polytypic for CD3, CD15 and CD20 in the lymphocyte immunophenotyping. IgG4/IgG ratio in plasma cells was < 40%. This analysis is consistent with RDD. In addition, the immunofluorescence evidenced membranous glomerulopathy in the immediate adjacent parenchyma, with focal granular deposits of mesangial and subendothelial distribution.

The patient remained asymptomatic until April 2017, when she complained of two weeks of progressive severe hypogastric pain, with a creatinine rise to 1.89 mg/dL. Abdominal CT scan evidenced right ureteral obstruction secondary to multiple retroperitoneal adenopathies up to 18 mm diameter, with accentuated hydronephrosis. A laparoscopic retroperitoneal lymphadenectomy was performed, and the biopsy informed a proliferative lesion composed of a dense monomorphic cellular infiltrate, again compatible with RDD.

After that, corticosteroids were started and the patient evolved favorably, without new episodes of extrinsic urinary compression nor reported complications. She remains in ambulatory medical controls with urologist and nephrologist.

Discussion

RDD is a very rare non-neoplastic condition, the majority seen in adolescents or young adults. The extranodal involvement is rare and the kidney is usually spared. In this case, we found renal parenchymal replacement and impairment of renal function. Foucar et al.[1] in 1990 described 10 cases with renal parenchyma replacement in their review. After that, about 14 cases have been published, where three of them had bilateral involvement of the renal parenchyma,[2–4] one also had testicular involvement[5] and two were associated with prostatic adenocarcinoma.[5] To the best of our knowledge, this represents the first case of renal RDD with secondary membranous glomerulopathy and the

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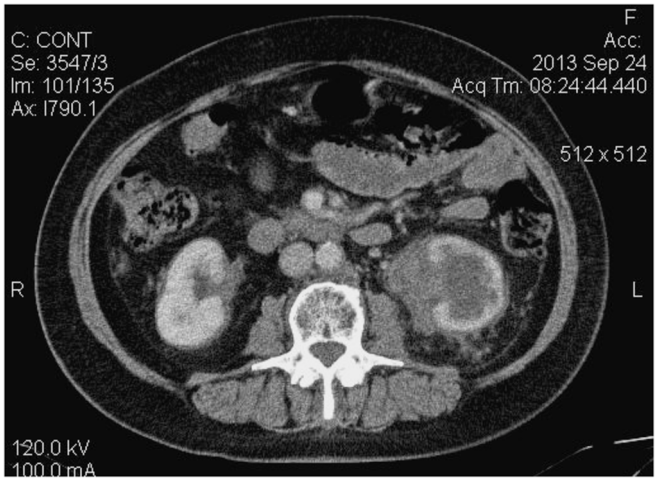

Fig. 1. CT that shows the mass of approximately 5 × 4.1 × 2.4 cm in relation to the left renal pelvis associated with moderate-accentuated hydronephrosis.

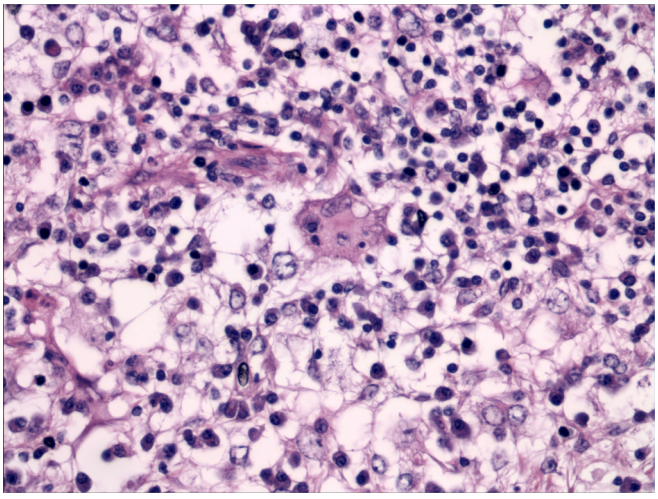

Fig. 2. Injury zone composed of a polymorphic inflammatory infiltrate, with lymphocytes, plasmocytes and large cells with histiocytic appearance, clear cytoplasm and also eosinophilic. At the center, one of the large histiocytes contains intracytoplasmic lymphocytes (emperipolesis). (H & E, 400×).

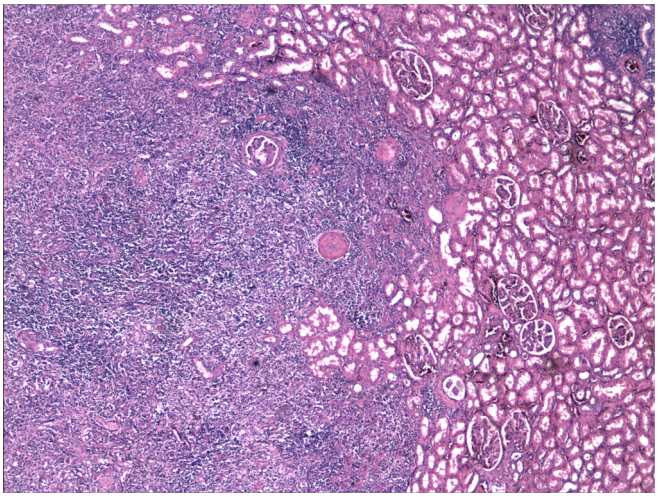

Fig. 3. Zone of renal cortex compromised by a proliferative lesion of neoplastic appearance composed of a dense and apparently monomorphic cellular infiltrate that replaces renal renal tissue in an intense manner and showing a definite limit with respect to the unaffected parenchyma (H & E, 50×).

third reported case of extranodal variant RDD presenting as a primary renal mass in an elderly patient without prior history of nodal variant disease.[3]

Conclusion

We present a very unusual form of presentation of this rare disease, which implies a worse prognosis. Patients with kidney involvement have worse outcomes, with up to 40% mortality. The survivors usually persist with chronic kidney disease(1). RDD should be considered in the differential diagnosis of benign lymphadenopathies with systemic involvement.

Informed consent

Permission for publication and informed consent from the patient was obtained.

Conflicts of interest

The authors declare no conflicts of interest.

Acknowledgment

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