

Retroperitoneal castleman disease mimicking lymph node spread from clear renal cell carcinoma. A case report

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ARTICLE INFO

Keywords:

Renal cell carcinoma
 Castleman disease
 Lymphadenectomy

ABSTRACT

Renal Cell Carcinoma (RCC) corresponds to 3% of the neoplasms in the adults. Surgery is the main mode of treatment, which can be associated to retroperitoneal lymphadenectomy in the presence of clinically tumor positive lymph nodes. Castleman Disease (CD) is a rare lymphoproliferative disorder, with little-known etiopathogenesis. It rarely affects the retroperitoneum. Thorax, neck, and abdomen are more frequently affected. Therefore, CD can simulate lymphatic spread from RCC to the retroperitoneum, also leading to a possible misdiagnosis, or diagnosis concerning a paraneoplastic syndrome due to RCC.

Introduction

Renal Cell Carcinoma (RCC) corresponds to 3% of all neoplasms in adult patients. The frequency of disease with histologically positive lymph nodes is up to ten percent (10%), a situation that is linked to lower survival rates.^{1,2}

The standard treatment option is surgery, as such as nephron-sparing surgery, like cryotherapy and radiofrequency ablation (types of minimally invasive percutaneous techniques), that can be utilized on RCC treatment in patients with nephropathies, bilateral neoplasms, or hereditary RCC, such as Von Hippel-Lindau (VHL) disease. Radiation therapy and chemotherapy are ineffective and immunotherapy has been the main choice of treatment of the metastatic renal carcinoma.^{1,2}

Castleman Disease (CD) is a rare disease, with heterogeneous presentations and different clinicopathological forms. Although it is of unknown etiology, this condition is associated with an action of cytokines and other immune mediators, such as interleukin 6, that plays an important role in the link between CD and epithelial malignancies. Castleman Disease can be classified in two main subtypes: Unicentric Castleman Disease (UCD) or Multicentric Castleman Disease (MCD). The retroperitoneum may be compromised in about 17% of the cases.^{3,4,5}

Case presentation

L.D, 60-year-old male, admitted to the hospital due to anemia, night sweats and weight loss. Computed Tomography scan detected splenomegaly, a renal mass on the left kidney and retroperitoneal lymph adenomegaly. At the physical examination, splenomegaly was found without evidence of a palpable renal mass. The predominant symptoms were fever, weight loss and night sweats. After multidisciplinary team discussion, the patient underwent a surgical procedure, which was a left side radical nephrectomy surgery with the resection extended to the spleen and retroperitoneal lymphadenectomy (Fig. 1).

The clinical evolution was satisfactory in the postoperative period. The patient received hospital discharge in the fifth day after admission. The histopathological diagnosis was Clear Cell Renal Cell Carcinoma (Fig. 2).

The largest diameter of the tumor was 10,8 cm, with a high-grade neoplasm with a big portion of tumoral necrosis and renal invasion at the renal pelvis. Of the 21 retroperitoneal lymph nodes that have been removed, all of them showed negative lymph node status for malignancy, but they had the presence of lymphoplasmacytic infiltrate. A posterior analysis of the lymph nodes, using the Immunohistochemistry

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<https://doi.org/10.1016/j.eucr.2020.101503>

Received 21 October 2020; Received in revised form 16 November 2020; Accepted 19 November 2020

Available online 30 November 2020

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Testing resulted on the confirmation of the absence of neoplastic entities at these lymph nodes. Angiofollicular lymph node hyperplasia was detected, with polyclonal plasmacytosis, confirming the diagnosis of Retroperitoneal Castleman Disease (Fig. 3).

Discussion

The RCC englobes a large and heterogeneous group of neoplasms, with uncertain clinical evolution. The fundamental prognostic factors, that are recognized as key determinants of the treatment, clinical evolution and overall survival of the patients are: clinicopathological staging, histological grade and performance status.²

The finding of positive lymph node status (involvement of lymph nodes, histologically positive for disease) or metastatic (M1 in the TNM staging system), implies a worse prognosis for the patient.^{1,2} Histologically positive nodes are present in 2–10% of the renal carcinomas, and the survival rates of these patients ranges from 11 to 35%.¹

There is indication for lymphadenectomy in RCC in the presence of a clinically tumoral lymph node disease, since retrospective studies demonstrated controversial results about the routine indications.¹ Thus, in the group of high-risk kidney neoplasms, the lymphadenectomies could benefit the taxes of response to the adjuvant immunotherapy treatment.^{1,2}

According to Ristau et al.,¹ the retroperitoneal lymphadenectomy procedures are realized when high-grade tumors are identified, in cases of left side location, on open surgeries and in the cases of radical nephrectomy, which involves the adrenal gland.

In this case report, the 60-year-old male patient presented with a renal mass located at the left kidney, that was larger than 10 cm and also with a retroperitoneal lymph adenomegaly. The patient was submitted to a radical nephrectomy extended to the spleen and adrenal gland, in addition to a retroperitoneal lymphadenectomy.

At the surgical procedure, enlarged lymph nodes were identified, forming a collection (or blocks) of lymph nodes, being interpreted as metastatic disease from the Renal Cell Carcinoma.

The histopathological report of the removed retroperitoneal lymph nodes demonstrated absence of neoplastic disease. Retroperitoneal

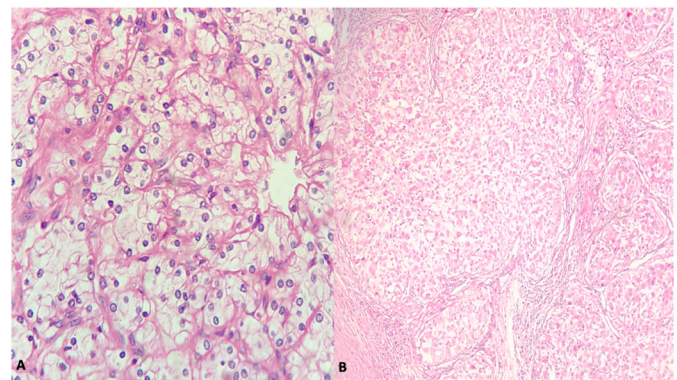


Fig. 2. A – Renal Clear Cell Carcinoma, HE 400 x. B – Renal Clear Cell Carcinoma with rhabdoid feature, HE 400 x.

Castleman Disease was confirmed by means of IHC (Immunohistochemistry), showing angiofollicular hyperplasia associated with polyclonal plasmacytosis, in spite of being negative for HHV-8 (Human herpes virus 8). The final diagnosis was Multicentric Idiopathic Castleman Disease.

Very commonly, CD patients have higher expression profiles of VEGF (Vascular Endothelial Growth Factor), in consequence of IL-6 and anemia stimulus, which are alterations frequently observed on RCC. Castleman Disease can generate splenomegaly, lymph adenomegaly and unspecific clinical signs, like fever, weight loss, and night sweats. These are common symptoms that RCC can also manifest, through paraneoplastic syndromes, in about 10–40% of the cases^{3,4,5}

Here in this study, by reason of the appearance of an extensive renal mass, retroperitoneal lymph adenomegaly and also the unspecific clinical symptoms in the patient, lymphatic tumoral dissemination and paraneoplastic syndrome were attributed, respectively, to this patient.

Conclusion

Besides being a rare clinicopathological entity, Castleman Disease

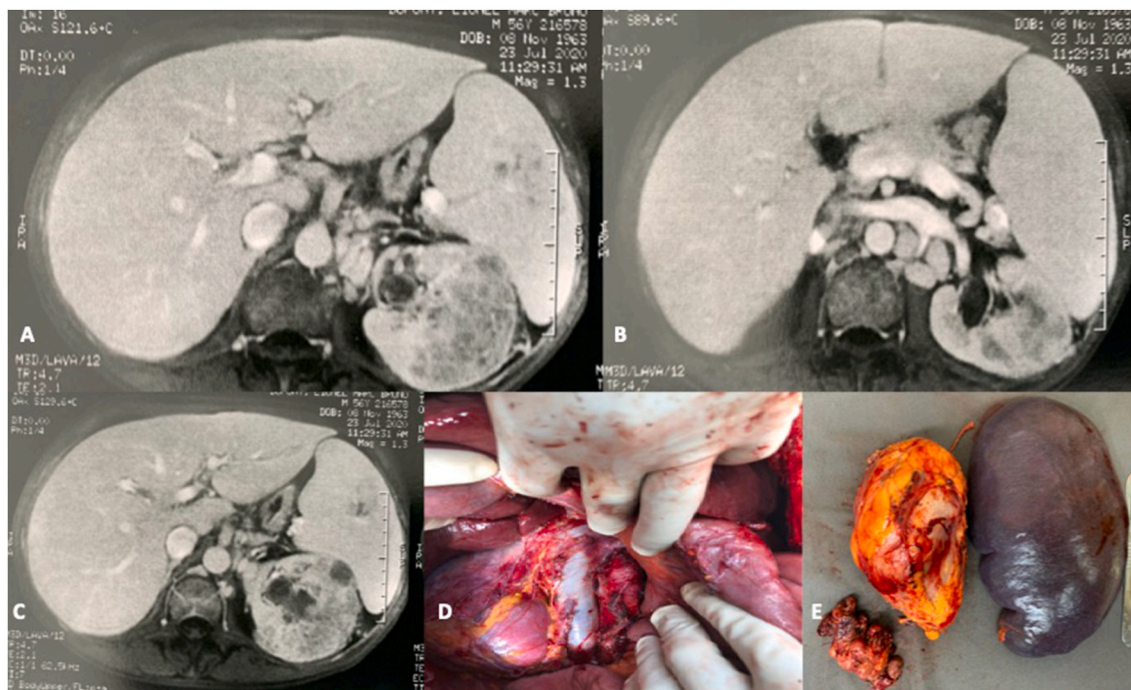


Fig. 1. A – CT scans demonstrate a left renal mass with extensive necrosis. B – Retroperitoneal and hilar lymph nodes. C – Left renal mass and splenomegaly. D – Retroperitoneal lymphadenectomy. E – Radical left nephrectomy, retroperitoneal and hilar lymph nodes and spleen.

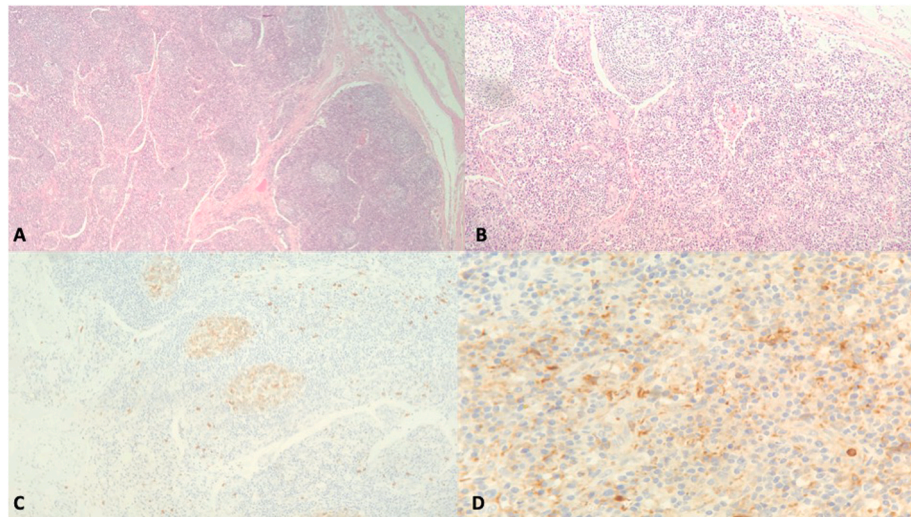


Fig. 3. A – Castleman disease and atypical lymphoid tissue. B – Presence of lymphocyte and plasmocytic infiltration. C – Immunohistochemistry demonstrating enlarge follicles. D – Immunohistochemistry demonstrating the plasmocytic infiltration.

can be a differential diagnosis of Renal Cell Carcinoma with regional lymphatic dissemination, once it simulates the presence of clinically active tumoral lymph nodes. The symptoms occasioned by CD, despite being unspecific, are very similar to those of paraneoplastic syndromes linked to RCC.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

1. Ristau BT, Manola J, Haas NB, et al. Retroperitoneal lymphadenectomy for high risk, nonmetastatic renal cell carcinoma: an analysis of the ASSURE (ECOG-ACRIN 2805) adjuvant trial. *J Urol.* 2018;199(1):53–59.
2. Barata PC, Rini BI. Treatment of renal cell carcinoma: current status and future directions. *Ca - Cancer J Clin.* 2017;67:507–524.
3. Wang HW, Pittaluga S, Jaffe ES. Multicentric Castleman disease: where are we now? *Semin Diagn Pathol.* 2016;33:294–306.
4. Fajgenbaum DC. Novel insights and therapeutic approaches in idiopathic multicentric Castleman disease. *Blood.* 2018;132:2323–2330.
5. Minemura H, Tanino Y, Ikeda K. Possible association of multicentric castleman's disease with autoimmune lymphoproliferative syndrome. *Biores Open Access.* 2018;7: 47–51.