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CASE REPORT Volume 5 Issue 3

# *Incidental Retroperitoneal Castleman's Disease Found in Patient with Renal Cell Carcinoma: a case report*

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## ABSTRACT

This report briefly discusses a case of retroperitoneal Castleman's disease in a 52 year old post-menopausal woman with renal cell carcinoma.

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**KEYWORDS**
Castleman's disease

## INTRODUCTION

Castleman's disease is a rare, benign, lymphoproliferative disorder first described in 1954 that affects less than 200,000 people in the United States.[1] While most cases involve the thorax, 15% of cases affect the abdomen or pelvis.[2] Castleman's disease is subdivided into two histopathological subtypes: hyaline-vascular variant or plasma cell variant.[1] The hyaline-vascular variant is associated with abnormal lymphoid follicular hyperplasia and increased interfollicular vascularity while the plasma cell variant is distinguished by sheets of mature plasma cells on histology.[3]

Castleman's disease is further divided clinically into unicentric or multicentric disease. Unicentric, or localized disease, is typically asymptomatic and has well circumscribed lesions with moderate to intense enhancement on imaging. Multicentric disease typically presents with systemic systems such as fever, night sweats, peripheral lymphadenopathy, or hepatosplenomegaly, and is often associated with human immunodeficiency virus (HIV) or human herpes virus-8 (HHV-8) infection, and is harder to distinguish from lymphoproliferative disorders.[1,3] In localized disease, 85- 90% of cases are hyaline-vascular variant.[1] Most are found incidentally on imaging as patients are asymptomatic or have few symptoms with vague complaints.[3] Radiological appearance is often nonspecific, so diagnosis must be made via biopsy or surgical resection.[4] We report a case of retroperitoneal unicentric Castleman's disease found incidentally in a patient with renal cell carcinoma.

## CASE REPORT

A 52 year old postmenopausal female presented to her primary care physician complaining of generalized abdominal tenderness and heavy vaginal bleeding for several weeks. The patient was given Provera 5 mg daily to control the bleeding. A computed tomography (CT) scan without contrast at this time showed a partially calcified right lower quadrant retroperitoneal mass of at least 5.2 cm concerning for malignancy, an enlarged uterus with possible leiomyoma, and a mass in the lower pole of the left kidney. CT-guided biopsy of the

retroperitoneal mass was non-diagnostic. Labs at this time showed normocytic anemia with a hemoglobin of 7.8 g/dL. A referral to urologic oncology and gynecologic oncology was made.

A retroperitoneal ultrasound showed a heterogeneous solid mass in the inferior left kidney. An endometrial biopsy showed complex endometrial hyperplasia with atypia. A repeat CT with and without contrast showed a solid enhancing mass of the inferior pole of the left kidney (5.1 x 4.4 x 4.3 cm) concerning for renal cell carcinoma and a lipomatous mass with irregular central calcification in the retroperitoneum of the right lower quadrant (soft tissue mass measuring 5.4 cm) unchanged from prior CT scan concerning for liposarcoma. Positron emission tomography (PET)/ CT imaging prior to surgery showed hypermetabolic activity in the retroperitoneal mass, mass of left kidney, thyroid, and the uterus. The centrally calcified soft tissue component of the retroperitoneal mass was hypermetabolic with peak standardized uptake value (SUV) of 5.1 concerning for malignant sarcoma. Decision was made to undergo robotic-assisted total laparoscopic hysterectomy, bilateral salpingo-oophorectomy, excision of retroperitoneal mass, and partial nephrectomy of left kidney. During surgery, partial left nephrectomy was converted to left radical nephrectomy. The surgery was otherwise uncomplicated. Specimens obtained included left kidney and ureter, miscellaneous renal fragments, 10 cm retroperitoneal mass, uterus, bilateral fallopian tubes, bilateral ovaries, cervix, and pelvic washings. Pathology included: left kidney mass showing stage 1b clear cell renal cell carcinoma limited to the kidney, uterus showing endometrial hyperplasia with atypia with multiple leiomyomas, and the retroperitoneal mass was diagnosed as hyaline vascular variant Castleman disease. Patient's post-operative period was uncomplicated.

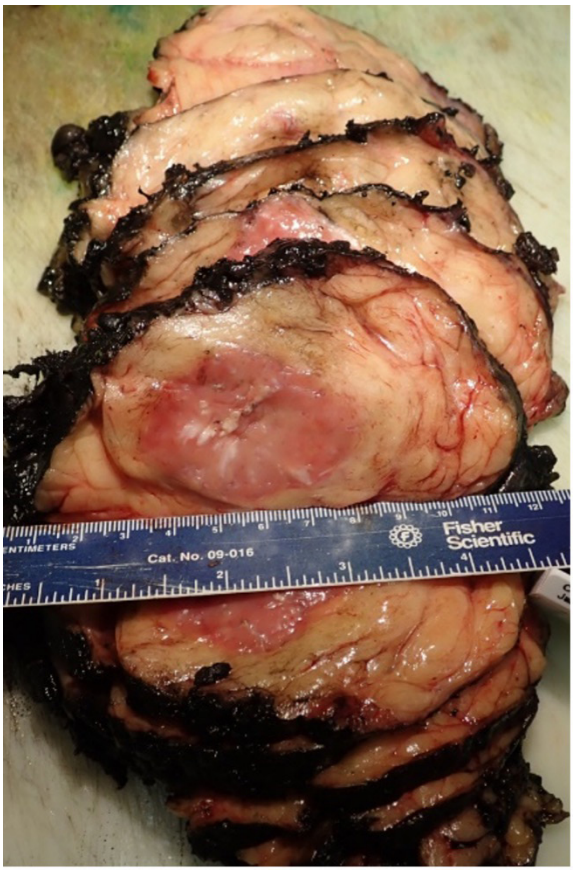


**IMAGE 1.** Gross picture of retroperitoneal mass with hyaline Castleman disease.

## DISCUSSION

While the etiology of Castleman's disease is unknown, development of the disease is thought to be attributed to an autoimmune process, immunodeficiency, chronic low-grade inflammation, or lymphoid hamartomatous hyperplasia.[2]

Our patient diagnosed with stage 1b renal cell carcinoma is in a state of chronic inflammation, which we hypothesize may have contributed to her disease process. Our patient presented with vague symptoms which is consistent with localized, unicentric Castleman's disease with hyaline-vascular variant.[3] However, due to our patient's unique situation of presenting with both Castleman's disease and renal cell carcinoma, it is unclear which etiology was causing her presenting symptoms. Our patient's repeat CT scan initially was concerning for a liposarcoma and further imaging with PET/ CT scan was concerning for sarcoma. Common differential diagnoses for Castleman's disease should include retroperitoneal sarcoma, lymphoma, desmoid tumor, carcinoid, or adenopathy.[1,2] Therefore, sarcoma is a common misdiagnosis for Castleman's disease and should be part of a differential diagnosis with retroperitoneal

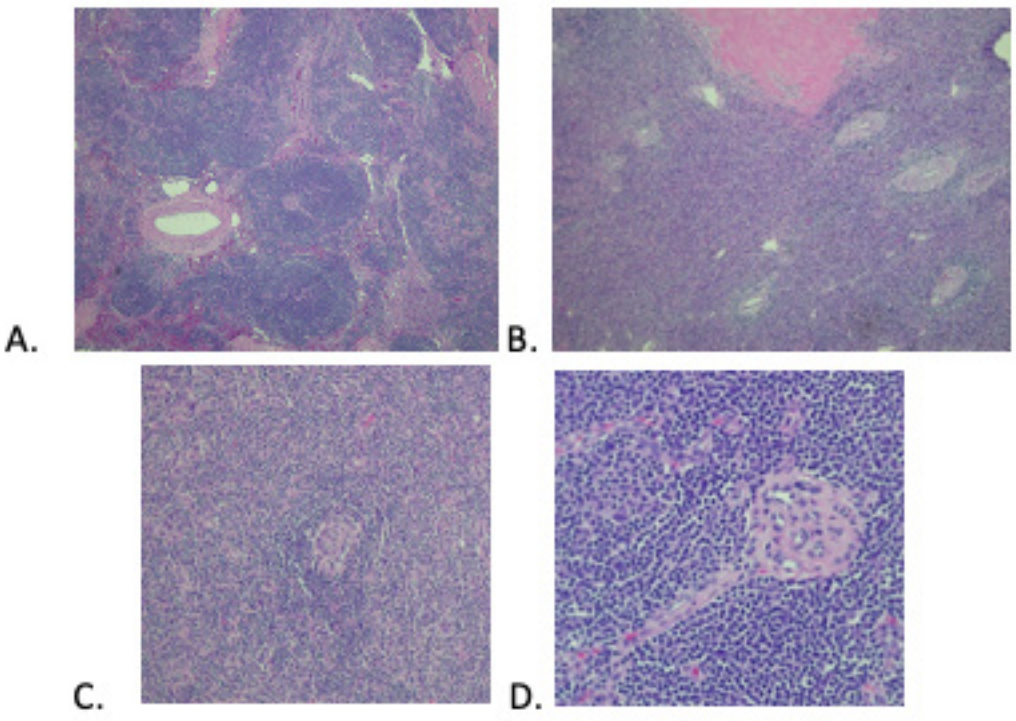


**IMAGE 2.** Microscopic picture of this case showing a) Depleted germinal center and expanded mantle zones, b) Expanded interfollicular area with hyalinized vessels, c) Regressed follicles with expanded interfollicular area, d) Depleted germinal center with penetrating vessels ("lollipop" appearance)

| Type of Castleman's disease (% of all cases) | Median age at diagnosis (years) | 5- year survival rate | % Plasma cell variant subtype | % hyaline vascular subtype |
|---|---|---|---|---|
| Unicentric/ Localized (47-81) | 35 | 91% | 9- 24% | 76-91% |
| Multicentric (19-53) | 55.5 | 27-90% | 77% | 19% |

**TABLE 1.** Comparison of Unicentric/ Localized and Multicentric Castleman's disease[3].

masses. In hyaline-vascular variant disease, 10% of masses are partially calcified, which is consistent with our patient who had centrally calcified soft tissue mass on CT scan.[2] While smaller masses may be homogeneous, larger masses are often heterogeneous due to central necrosis.[3] PET imaging of Castleman's disease often demonstrates an SUV of fluorodeoxyglucose (FDG) in a range lower than seen in patients with lymphoma, which may be able to help differentiate between benign Castleman's disease and malignant lymphoma.[4] Ngeow et al. states that FDG uptake greater than 10 SUV is predictive of an aggressive B-cell lineage or suggestive of more aggressive histological components[5]. Our patient had an SUV of 5.1, which is lower than this predictive value.

Our patient was diagnosed via tissue biopsy after full surgical resection of the peritoneal mass. Due to the locally invasive nature of the tumor, patient's symptoms respond well to surgical resection.[1] Complete surgical resection of the mass is considered curative, and recurrence has only been described after incomplete surgical resections.[6] Therefore, our patient was treated according to the standard of care. Five year survival rate is nearly 100% in full surgical resection.[6] In multicentric disease, systemic chemotherapy is required and mean survival is fourteen to thirty months.[1] Our patient will have repeat imaging with a CT of chest, abdomen, and pelvis three months after surgery as well as continual follow up with a medical oncologist regarding her renal cell carcinoma.

## AUTHOR AFFILIATION

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## REFERENCES

1. Madan R, Chen JH, Trotman-Dickenson B, Jacobson F, Hunsaker A. The spectrum of Castleman's disease: mimics, radiologic pathologic correlation and role of imaging in patient management. Eur J Radiol. 2012;81(1):123-31.
2. Bonekamp D, Hruban RH, Fishman EK. The great mimickers: Castleman disease. Semin Ultrasound CT MR. 2014;35(3):263-71.
3. Jongsma TE, Verburg RJ, Geelhoed-Duijvestijn PH. Castleman's disease: A rare lymphoproliferative disorder. Eur J Intern Med. 2007;18(2):87-9.
4. Stephen P. Murphy MAN, Mark W. Karwal. FDG-PET Appearance of Pelvic Castleman's Disease. Journal of Nuclear Medicine. 1997;38:2
5. Ngeow JYY, Quek RHH, Ng DCE, Hee SW, Tao M, Lim LC, et al. High SUV uptake on FDG–PET/CT predicts for an aggressive B-cell lymphoma in a prospective study of primary FDG–PET/CT staging in lymphoma. Annals of Oncology. 2009;20(9):1543-7.
6. Pascal Bucher GC, Guillaume Zufferey, Frederic Ris, Olivier Huber, Philippe Morel. Surgical management of abdominal and retroperitoneal Castleman's disease. World Journal of Surgical Oncology. 2005.