



Case Report

Retroperitoneal pararenal castleman's disease: a case report

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ABSTRACT

Castleman's disease is a rare lymphoproliferative disorder of unknown etiology. We report a 28 years old woman with solitary Castleman's disease in the left pararenal space. This case was diagnosed preoperatively as renal cell carcinoma. The patient underwent a radical nephrectomy with dissection of pararenal mass. Histopathological examination of the surgically resected specimen showed the hyaline vascular type of Castleman's disease. A preoperative diagnosis of Castleman's disease is difficult; therefore, a surgical resection and histopathological evaluation can provide an accurate diagnosis of tumor. Taking this case into consideration, we suggest that Castleman's disease should be included in the differential diagnosis of renal tumors.

INTRODUCTION

Castleman's disease (CD), or angiofollicular lymph node hyperplasia, is a relatively rare disorder characterized by the benign proliferation of lymphoid tissue. It was first described by Castleman et al 1 in 1956. Castleman's disease arises mostly in the mediastinum. Some cases of non-mediastinal origins have been reported, but pararenal tumor are extremely rare. Castleman's disease has been classified as unicentric (localized) forms and multicentric (generalized) form based on clinical and radiological findings. Based on histopathological findings, it is classified into hyaline vascular type, plasmacytic and mixed cellular types. The hyaline vascular type is more common in the localized CD (LCD) type and the plasma cell type which is more common in multicentric CD (MCD) type. We report herein an unusual case of Castleman's disease in pararenal fossa of a 28 years old lady.

CASE REPORT

A 28 year-old lady was admitted to our hospital with complains of left middle abdominal pain. On admission, she was asymptomatic and physical examination revealed ill-defined mass in the left loin, with no surrounding tenderness. Ultrasonography of abdomen revealed a 75 x 52 mm sized hypoechoic mass at lower medial aspect of left kidney. Contrast enhanced CT scan showed a well-defined, intensely enhancing mass located in lower anterior medial aspect of the left kidney measuring approximately 75 x 52 mm in size (fig. 1). One of the differential diagnoses, of the CT imaging findings, was renal cell carcinoma or retroperitoneal lymphoma. Patient underwent radical nephrectomy. Intraoperative the mass was located in the retroperitoneal region, inside the perinephric fat at the lower anterior medial side of the left kidney. Its capsule was adherent to the lower pole of the kidney. There was no evidence of invasion into the adjacent tissues.

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Gross examination showed a well encapsulated tumor mass measuring 7.5 x 7.0 x 5.0 cm. Cut surface was solid, grayish and lobular with grey brown areas in the center (fig.2)

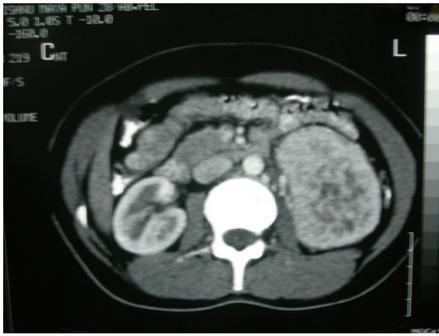


Figure 1: Contrast enhanced CT showing well-defined enhancing mass at left lower pole of the kidney measuring 7.0 cm in diameter



Figure 2: Macroscopic view of the resected left kidney (upper) attached with tumor mass (lower)

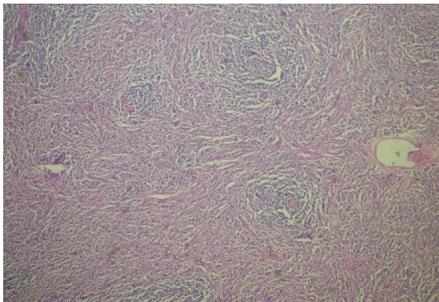


Figure 3: Hyaline vascular type of CD. Note the concentric rings of small lymphocytes surrounding atrophic follicles (HE stain, X40).

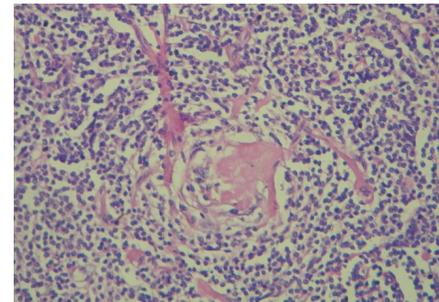


Figure 4: Hyaline vascular type of CD. The hyalinized vessel is entering the follicles (HE stain, X100).

Microscopically, the tumor had completely encapsulated huge lymph node and revealed diffuse nodular expansion with regression germinal centre composed of large cells with vesicular nuclei. Some of the follicles showed vascular proliferation and hyalinization of the germinal center. Interfollicular area contained large number of lymphocytes and showed capillary proliferation (fig.3A&B). There were also seen ten small para-aortic lymph nodes which showed features of reactive lymphoid hyperplasia. Multiple sections from the renal parenchyma were unremarkable. These histologic findings were consistent with retroperitoneal hyaline vascular type Castleman's disease.

DISCUSSION

Castleman's disease is a rare lymphoid tissue described in Castleman in 1954.¹ It is also known as angiofollicular hyperplasia, giant lymph node hyperplasia and lymphoid hamartoma or follicular reticuloma. Castleman's disease arises mostly in the mediastinum and about 70 % of the reported cases tend to be of mediastinum origin. The superficial nodal groups constitute 20% of cases. Retroperitoneal and pararenal localization are very rare and have been reported to account for 7% and 2% of cases, respectively.² Our patient had Castleman's disease just around the kidney.

The etiology of Castleman's disease is unclear, although several immunological mechanisms have been proposed including overproduction of interleukin-6 and human herpes virus type 8 infection.³

Two histological variants of the Castleman's disease have been identified: the hyaline vascular type, comprising 80-90% of the cases and plasma cell type comprising the remaining 10-20%.⁴ Clinically also two types have been described either unicentric localized form or multicentric generalized form. Unicentric CD usually presents as an isolated benign lymphoproliferative disorder in young adults (average 33 years), mostly asymptomatic and identified incidentally as an abnormality on imaging studies. Multicentric CD usually presents in the sixth decades. It manifests as a systemic disease with generalized peripheral lymphadenopathy, hepatosplenomegaly, frequent fever and has a poor prognosis.

Microscopically, two major histopathologic categories have been described: the hyaline vascular (HV) and the plasma cell (PC) variants. The hyaline vascular type shows numerous small involuted follicular centers in enlarged lymphoid tissue. The follicles show marked vascular proliferation and hyalinization of their abnormal germinal centers. There is a tight concentric layering of lymphocytes at the periphery of

the follicles resulting in an onion-skin appearance. The PC type is characterized by diffuse plasma cell proliferation in the interfollicular tissue, while hyaline vascular changes in the follicles are inconspicuous or absent.

Unicentric CD of either the hyaline vascular type or plasma cell variants is almost universally cured after resection of the lymph nodes involved and has not been associated with increased mortality.⁵

Diagnostic imaging methods, such as ultrasound and CT generally cannot differentiate CD from other diseases due to the lack of tumor-specific signs, but these examinations do enable us to locate the exact position of a tumor. Only a surgical resection and a histological evaluation can provide us with an accurate diagnosis of this tumor.

CONCLUSION

Retroperitoneal hyaline vascular type of Castleman's

disease although rare should be included in the differential diagnosis of retroperitoneal mass like other malignant renal tumors. A preoperative diagnosis of CD is difficult; therefore, a surgical resection and a histological evaluation can provide an accurate diagnosis of this tumor.

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