CASE REPORT

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Castleman's disease mimicking right adrenal neoplasm: A case report

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Castleman's disease usually manifests as a solitary mediastinal tumor and only rarely as an isolated retroperitoneal mass. This disorder is often undiagnosed or misdiagnosed. Thus, only very few patients have been reported and little information is available in the literature. The definitive diagnosis is based on postoperative pathological findings. We report a case of a 57-year-old female with a Castleman's tumor located superomedial to the upper pole of the right kidney that mimicked an adrenal neoplasm. The mass was surgically resected, and the histopathological diagnosis of the resected tissue was hyaline-vascular type of Castleman's disease. Although retroperitoneal Castleman's disease is rare, it should be considered in the differential diagnosis of retroperitoneal masses.

Key words: Castleman's disease, lymphoproliferative disorder, retroperitoneal neoplasms

Sağ adrenal tümörü taklit eden Castleman hastalığı: Olgu sunumu

Castleman's hastalığı, genelde mediastende görülür. İzole retroperitoneal kitle şeklinde görülmesi oldukça nadirdir. Genelde teşhisi konulamaz veya yanlış konulur. Kesin tanıda patoloji önemlidir. Bu konuda literatürde az sayıda vaka vardır. Sunduğumuz olgu, böbrek üstü kitlesi izlenimini veren, sağ böbreğin üst iç kısmında yer alan, 57 yaşında olan bir bayan hastadır. Eksize edilen kitlenin patolojisi vasküler tip Castleman's hastalığı olarak gelmiştir. Sonuç olarak, retroperitoneal bölgede Castleman's hastalığı nadir görülse de, ayırıcı tanıda akılda tutulmalıdır.

Anahtar kelimeler: Castleman hastalığı, lenfoproliferatif hastalık, retroperitoneal neoplaziler

INTRODUCTION

Castleman's disease (CD) is an uncommon lymphoproliferative disease that was first described in 1956 (1). The etiology and pathogenesis are still under debate. Although CD may occur anywhere along the lymphatic chain, the mediastinum is the most common location (70%). Extrathoracic sites such as the neck, axilla, pelvis, and retroperitoneum have been reported less frequently (2). The disease has been subdivided into two clinical patterns as unicentric and multicentric forms. It is considered benign in its localized form, but aggressive in the multicentric type (3). Because of the variability of clinical presentation, selection of the appropriate therapeutic approach remains unclear. We present a rare case of unicentric CD of the hyaline-vascular type with a right suprarenal location that mimicked an adrenal neoplasm.

CASE REPORT

A 57-year-old female patient with a two-month history of constant and dull left flank pain was investigated by ultrasonography (USG), computed tomography (CT) scan and magnetic resonance

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imaging (MRI) of the abdomen, which disclosed a retroperitoneal pararenal mass (8x7x6 cm) with heterogeneous imaging characteristics (Figure 1). The patient's history was not relevant, and she had not been operated previously. Moreover, there was no marked family history of cancer or any other specific disease.

She had no systemic symptoms such as sweating, weight loss, fatigue, or fever. On the physical examination, vital signs were all within normal limits. No palpable mass was found in the abdomen. There was no superficial lymphadenopathy or organomegaly. Further findings, including serum cortisol level, urine vanillylmandelic acid and serum aldosterone, were all within normal range. Based on the hypervascularity of the mass and the lack of specific signs in the imaging investigation, lymphoma, or mesenchymal or vascular tumors were considered as probable diagnoses, and surgical exploration was planned. Laparotomy revealed a solid oval mass situated in the retroperitoneum, adjacent to the right adrenal tissue and renal hilus.

The retroperitoneal lesion was removed in its entirety. Macroscopically, the mass was 10.5x7.5x6 cm with a firm, gray-white lesion, measuring 5.5x5x3.5 cm in its cut surface. The histological examination revealed broad bands of hyalinization that entrapped lymphoid follicles. Most of the follicles had hyalinized hypervascular germinal centers (Figures 2, 3). There were plasma cells and scattered eosinophils. The lesion had expansile borders rather than being infiltrative. No sign of an epithelial tumor was observed, and Hodgkin lymphoma was excluded in the differential diagnosis both with the absence of Reed-Sternberg cells and variants and the negativity of CD30 and CD15. With the above-mentioned histopathological findings, the diagnosis was established as "hyaline type" CD. The patient was discharged on the 5th day uneventfully.

DISCUSSION

Castleman's disease (CD) can be detected incidentally through the discovery of a slow-growing mass or by presentation with general symptoms, such as fever, failure to thrive or weight loss. The duration of symptoms or lymphadenopathy may vary from a few weeks to many months. The unicentric hyaline-vascular type of CD associated with systemic symptoms is rarely reported (<10%). The most commonly described (77–91%) symptoms in the literature are a localized and asymptomatic mass (4), as shown by our patient.

Treatment of localized CD usually involves resection, with excellent long-term results (5). CD patients mainly have lymphadenectasis in a single location, no clinical symptoms and normal laboratory results, with a majority of hyaline-vascular type. Although CT is helpful for the diagnosis of CD, the final diagnosis depends on pathologic examination. Localized CD patients can live long without recurrence after complete surgical resection of the tumor.

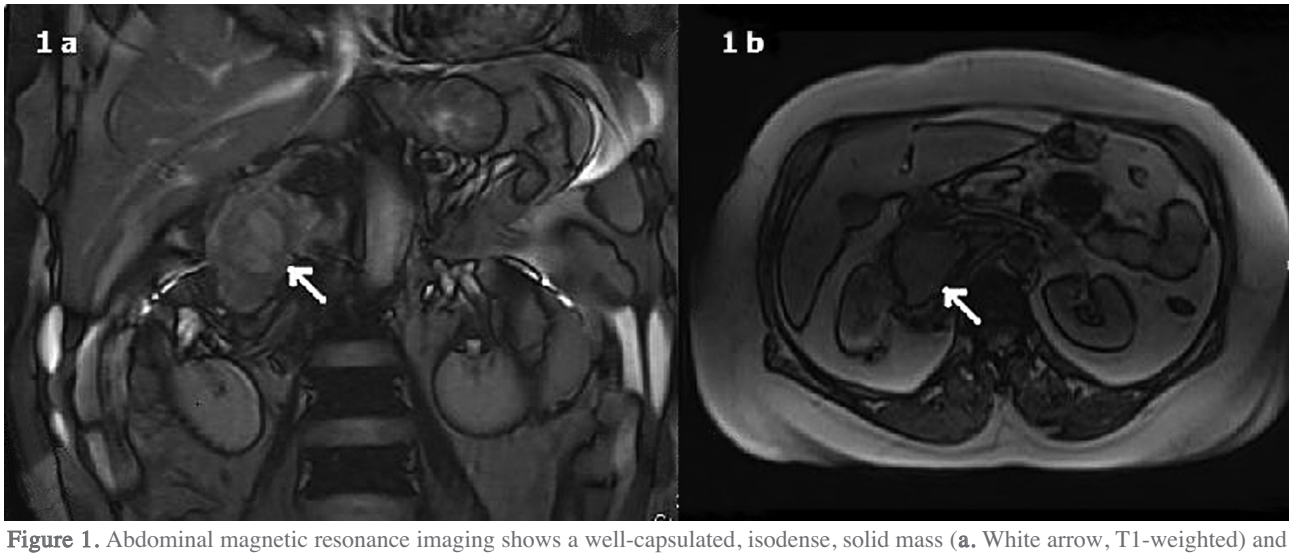

Figure 1. Abdominal magnetic resonance imaging shows a well-capsulated, isodense, solid mass (**a.** White arrow, T1-weighted) and heterogeneous signal characteristics, i.e., a central hypodense area within the mass (**b.** White arrow T2-weighted).

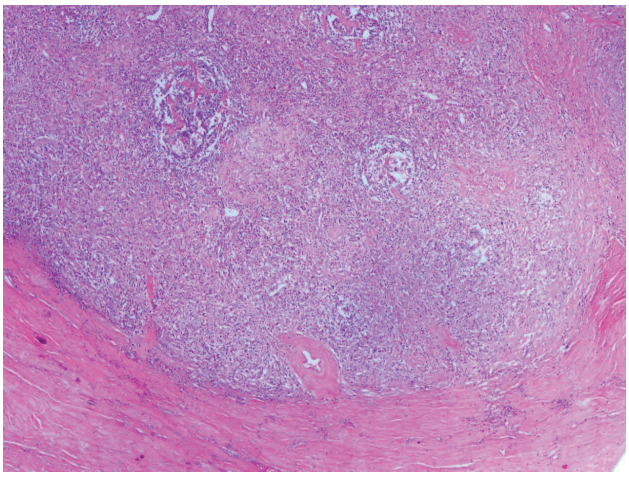

Figure 2. Lymphoid tissue surrounded by fibrosis and containing hypervascular hyalinized germinal centers (HE X40).

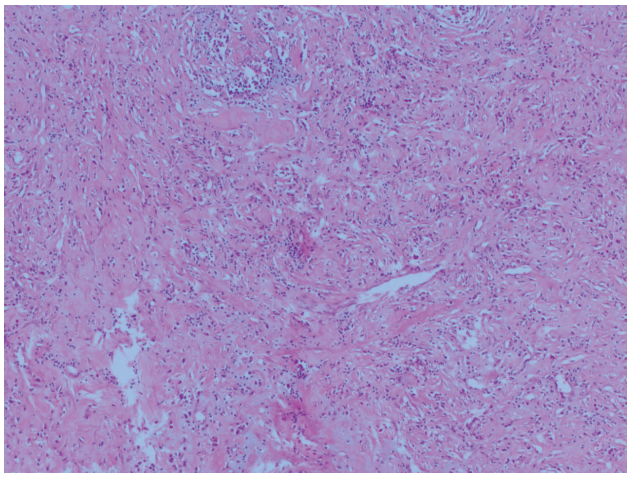

Figure 3. The microphotograph represents a broadly hyalinized part of the lesion with an atrophic lymphoid follicle (HE X100).

Based on the histological characteristics, Keller et al. (2) classified this disease into two variants: the hyaline-vascular type and the plasma-cell type. Surgical excision is both the diagnostic and the curative method for management of the disease.

Localized disease manifests as a solitary mass, which may be well circumscribed or infiltrative. It is associated with lymphadenopathy confined to one lymph node or nodal area, and usually follows a benign course. Multicentric disease carries a worse prognosis, and subsequent infection or malignancy may lead to death. Generally, complete surgical resection is sufficient to cure localized CD (5,6).

Unlike patients with localized CD, symptomatic patients with multicentric CD require systemic therapy, and several available alternatives matched to their needs and overall condition, including chemotherapy, steroids and antiviral therapy. Suppression of the immune response is the most important component of therapy (7). The use of prednisone or other steroids will frequently and promptly improve symptoms. Nishimoto et al. (8) reported the use of cilizumab, a humanized monoclonal antibody of the human interleukin (IL)-6 receptor, in a series of 28 human immunodeficiency virus (HIV)-negative patients. Recognition of the contribution of IL-6 inhibition to the development of multicentric CD has led to an attempt to block IL-6 or its effects directly.

Although this is an exceedingly rare condition, a diagnosis of localized CD could be considered whenever the patient has a solid, heterogeneous mass in the retroperitoneal area. An understanding of the clinical features of this disease would help prevent unnecessarily risky or extensive operations for the removal of these benign masses. The lesions usually grow slowly, and for patients with extensive comorbidities or questionable health status, conservative management should be considered (9).

An asymptomatic retroperitoneal mass in a young adult always raises the suspicion of a malignant tumor, but it is necessary to consider hyaline-vascular type CD prominently in the differential diagnosis when the arborizing pattern of calcification is seen.

In conclusion, unicentric CD is manifested in the form of benign, painless, slow lymph node enlargement that is generally asymptomatic. Complete surgical removal is recommended as a course of curative treatment. The multicentric form of CD exhibits a progressive clinical course with potential for malignancy. There is currently no standard therapy for multicentric CD. CD should be included in the differential diagnosis of any solitary, heterogeneous and hypervascular retroperitoneal mass.

REFERENCES

1. Castleman B, Towne VW. Case records of the Massachusetts General Hospital: Case No. 40231. N Engl J Med 1954; 251: 1001-5.

2. Keller AR, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. Cancer 1972; 29: 670-83.

3. Ye B, Gao SG, Li W, et al. A retrospective study of unicentric and multicentric Castleman's disease: a report of 52 patients. Med Oncol 2009 Nov 24 [Epub ahead of print].
4. Chen JH, Yu CY, Pai CY, et al. Castleman's disease in the left upper retroperitoneal space mimicking an adrenal neoplasm: report of a case and literature review. Jpn J Clin Oncol 2005; 35: 353-6.
5. Bowne WB, Lewis JJ, Filippa DA, et al. The management of unicentric and multicentric Castleman's disease: a report of 16 cases and a review of the literature. Cancer 1999; 85: 706-17.
6. Bucher P, Chassot G, Zufferey G, et al. Surgical management of abdominal and retroperitoneal Castleman's disease. World J Surg Oncol 2005; 3: 33.
7. Fowler A, Collins L, de Ruiter A, et al. Multicentric Castleman's disease in a patient with HIV. Int J STD AIDS 2006; 17: 63-4.
8. Nishimoto N, Kanakura Y, Aozasa K, et al. Humanized antiinterleukin-6 receptor antibody treatment of multicentric Castleman disease. Blood 2005; 106: 2627-32.
9. Sadamoto Y, Abe Y, Higuchi K, et al. Retroperitoneal Castleman's disease of the hyaline vascular type presenting arborizing calcification. Intern Med 1998; 37: 691-3.