Castleman’s Disease in the Left Upper Retroperitoneal Space Mimicking an Adrenal Neoplasm: Report of a Case and Literature Review

Jia-Hui Chen1,4, Chih-Yung Yu2, Chien-Yu Pai3, De-Chuan Chan1, Chung-Jueng Chen1, Jyh-Cherng Yu1 and Yao-Chi Liu1

1Division of General Surgery, Department of Surgery, 2Department of Radiology and 3Department of Pathology, Tri-Service General Hospital, National Defense Medical Center, Taipei and 4Department of Surgery, Hualien Armed Forces General Hospital, Hualien, Taiwan, Republic of China

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Castleman’s disease is a rare disorder characterized by benign proliferation of lymphoid tissue. Most cases occur as a mediastinal mass, although extrathoracic involvement including nodal and extranodal locations has been reported. The left suprarenal location of this localized disease may be mistaken for an adrenal tumor. We report a case of a 51-year-old woman with a Castleman’s tumor located superomedial to the upper pole of the left kidney that mimicked an adrenal neoplasm.

Key words: Castleman’s disease – giant lymph node hyperplasia – mimicking adrenal neoplasm

INTRODUCTION

Castleman’s disease (CD) is an uncommon and poorly understood disorder of lymph node hyperplasia with unknown etiology that was described first by Castleman in 1954 (1, 2). CD is classified into two clinical subtypes: a localized (or unicentric) subtype and a multicentric subtype. 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 (3), and usually follows a benign course. Multicentric disease carries a worse prognosis, and subsequent infection or malignancy may lead to death (3).

CD can also be classified into two major histological subtypes: a hyaline-vascular subtype and a plasma cell subtype. The hyaline-vascular subtype is more common and contains numerous regressively transformed follicles and associated vascular proliferation. The plasma cell type contains hyperplastic follicles and marked plasma cell proliferation in the interfollicular region. A mixed form, also called the hyaline-vascular–plasma cell subtype, is uncommon (4).

CD may occur anywhere along the lymphatic chain, although the mediastinum is the most common location (70%). Extrathoracic sites have been reported in the neck, axilla, pelvis and retroperitoneum (4). We present a rare case of unicentric CD of the hyaline-vascular type with a left suprarenal location that mimicked an adrenal neoplasm.

CASE REPORT

A 51-year-old woman was hospitalized with a 2 week history of poor appetite and a loss of body weight of ~6 kg. Her history included total abdominal hysterectomy and left salpingoopherectomy. She was otherwise well, had no pertinent medical history and was not taking any medication. Physical examination revealed no peripheral lymphadenopathy and no significant abnormality. Laboratory data and tumor markers were within the normal limits. Ultrasonography demonstrated a well-circumscribed hypoechoic mass over the medial aspect of the left suprarenal area. Post-contrast computed tomography (CT) revealed a well-defined enhancing mass (size: 5.2 cm × 4.7 cm × 5.6 cm) with multiple small areas of central low attenuation at the medial aspect of the left suprarenal region (Fig. 1). Plain magnetic resonance imaging (MRI) demonstrated a hypo-intense on T1-weighted image and mild hyper-intense on HASTE T2-weighted image with multiple small foci of very high signal intensity (Fig. 2). This tumor did not display signal dropout on the out of phase spoiled gradient-echo (SGE) T1-weighted image, which confirmed a lack of fat component within the mass. The contrast-enhanced fat saturation T1-weighted image also disclosed an enhancing pattern similar to that shown by CT. During laparotomy, a yellow mass was located below the left adrenal gland, upon the left kidney, and lateral to the abdominal aorta. Extensive resection of the mass was performed and one well-capsulated mass in size and 84 g in weight was resected (Fig. 3). Pathological examination demonstrated characteristics of the hyaline-vascular type of CD (Fig. 4). The patient was not given any radiation or chemotherapy, but was...
advised that low-dose radiotherapy would be considered for any further recurrence. She has had no clinical or radiological recurrence and remains free of the disease after 2 years.

DISCUSSION

Disorders of the adrenal gland result in classic endocrine syndromes such as Cushing syndrome, hyperaldosteronism and pheochromocytoma. In addition, tumors of the adrenal glands may present with abdominal pain or as an abdominal mass. The diagnosis of these disorders requires careful endocrine evaluation and, in many patients, adrenal imaging studies are required to define adrenal anatomy. In our patients, pulse, blood pressure, renal function tests, serum cortisol, electrolytes and urinary catecholamines were all normal. Preliminary diagnosis was therefore non-functioning adrenal tumor. However, the tumor located below the left adrenal gland during operation and the diagnosis of CD depended on the histopathological examination.

CD is a poorly understood lymphoproliferative disease that occurs mainly in young, otherwise healthy patients, although the age may range from 8 to 66 years; males and females are equally affected (4). The etiology remains unclear, although several immunological mechanisms have been proposed, including overproduction of interleukin-6 and human herpes virus type 8 infection (5).

CD may occur in almost any area where lymphoid tissue is normally found (4). Of the 400 reported cases (6), 70% involved the thorax, 40% the neck, 12% the abdomen and 4% the axilla. Most intra-abdominal lesions are located in the pelvic, mesenteric and perinephric regions (7), but such lesions have been described throughout the abdomen. To our knowledge, the world literature has described only seven cases of CD in the upper urinary tract (8–11). Debatin et al. reported that CD might originate from the lymphoid tissue around the adrenal gland and might extend subsequently into the adrenal gland itself (12).
CD is infrequently associated with various immunological abnormalities or with the subsequent development of a malignancy such as Kaposi's sarcoma, malignant lymphoma or plasmacytoma. Three histological variants (hyaline-vascular, plasma cell and mixed) and two clinical types (unicentric and multicentric) have been described (3,4). The hyaline-vascular variety accounts for up to 90% of CD and is usually asymptomatic; the plasma cell subtype is less common, and ~50% of these patients experience anemia, fever, fatigue, hyperglobulinemia and hypoalbuminemia (13).

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 lymphoadenopathy may vary from a few weeks to many months. The unicentric hyaline-vascular type of CD associated with systemic symptoms is rarely reported (<10%) (4,14). The most commonly described (77–91%) symptoms in the literature are a localized (4,14) and asymptomatic mass (2,4), as shown by our patient.

Common laboratory anomalies include anemia, hypalbuminemia, polyclonal gammopathy, elevated erythrocyte sedimentation rate or C-reactive protein concentration, and proteinuria (15). Diagnostic imaging methods such as ultrasound and CT or MRI cannot identify CD because of the lack of tumor-specific signs, but these tests yield important information about the exact tumor location (13). Gallium scintigraphy is also considered a sensitive tool for diagnosis and detection of the hyaline-vascular type of CD, but its utility in detecting the plasma cell variant is debated (16).

The clinical manifestations and radiological findings in our patients include poor appetite, weight loss and a solitary mass over the medial aspect of the left suprarenal region abutting the adrenal gland. Only surgical resection and conventional histological evaluation can give an accurate characterization of this tumor.

Treatment of localized CD usually involves resection, with excellent long-term results (3). Radiotherapy has also been reported to be effective in some patients with unicentric disease and is considered a treatment option for patients who are poor surgical candidates or have undergone incomplete resection (5). Patients with multicentric CD do not benefit from surgical treatment and should be candidates for steroid treatment, with or without chemotherapy (3,5). Our patient received surgical resection and was not given any radiation or chemotherapy. Low-dose radiotherapy was suggested for any further recurrence, and no clinical or radiological recurrence was noted for 2 years.

This case appears to be unique. The location of CD over the left adrenal region in patients with the unicentric subtype has not been documented previously. Diagnosing CD can be difficult, given the non-specificity of its systemic manifestations, lack of knowledge on its etiology, and the need for a histopathological examination to confirm the diagnosis. Further pre-operative imaging studies including gallium scintigraphy may be helpful in diagnosing CD.

References