CASE REPORT

Retroperitoneal Castleman’s Disease of the Hyaline Vascular Type Presenting Arborizing Calcification

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Castleman’s disease (CD) usually manifests as a solitary mediastinal tumor and only rarely as an isolated retroperitoneal mass. In the latter instances it is difficult to distinguish radiographically from other retroperitoneal masses. We report a 22-year-old female patient with retroperitoneal CD of the hyaline vascular type presenting with arborizing calcification. This characteristic calcification pattern is considered unique to CD, and is useful in diagnosis when present. (Internal Medicine 37: 691-693, 1998)

Key words: retroperitoneal tumor, calcification

Introduction

In 1954 Castleman first described an asymptomatic benign hyperplastic lymph node resembling a thymoma (1). Presently, this condition is known as Castleman’s disease (CD), angiofollicular lymph node hyperplasia, or giant lymph node hyperplasia. Two histologic types are recognized: hyaline vascular (HV) and plasma cell (PC). The hyaline vascular type accounts for 90% of cases, and usually is solitary and asymptomatic except for possible compression of adjacent structures. On the other hand, patients with PC type typically have fever, general fatigue, anemia and polyclonal hypergamma-globulinemia. CD usually occurs within the mediastinum but occasionally in the retroperitoneum (2). Morishita et al reviewed 59 cases of retroperitoneal CD in Japan: on 1992 (3). CD of HV type usually has no distinctive clinical or radiographic features, making it difficult to distinguish from other retroperitoneal tumors (4-7). We report a retroperitoneal case showing arborizing calcification, which is not typically found but is diagnostic for HV type CD when present (8-10).

Case Report

A 22-year-old female was admitted to our hospital because of pain in the right hypochondrium due to right urinary tract infection (UTI). On an abdominal radiograph, an arborizing calcification measuring 7×6 cm was noted in the left upper abdomen (Fig. 1). On physical examination, a mass was distinctly palpable in the same place. After the treatment of UTI, the patient presented no symptoms and routine laboratory data were normal.

Gallium scintigraphy showed no abnormal uptake. Ultrasonography revealed a large solid mass with mixed echogenicity; the highly echogenic area with acoustic shadowing was thought to represent the calcification. On computed tomography (CT), the mass was solid and relatively well demarcated, and displayed arborizing calcification; it was located in the retroperitoneum ventral and inferior to the left kidney (Fig. 2). An enlarged lymph node was seen in the left lower paraaortic region. T1-weighted magnetic resonance imaging showed the signal from the mass to be heterogeneously hypointense relative to that from the liver. T2-weighted imaging demonstrated heterogeneously hyperintense signal characteristics within the mass; the margin separating the mass from the surrounding soft tissue was not uniformly distinct. Angiographically the mass was hypervascular, showing a dense capillary blush supplied by the first jejunal artery.

At laparotomy an encapsulated tumor was found in the left pararenal region and an enlarged paraaortic lymph node was also removed. Dissection of the tumor was difficult due to thick vascular fibrous adhesions between the tumor capsule and other retroperitoneal structures. The tumor measured 9×6×6 cm and weighed 165 g (Fig. 3). The lymph node measured 5×3×3 cm. Microscopically, the tumor and lymph node showed hyperplastic lymphoid tissue with numerous follicles, and vascular proliferation in interfollicular areas (Fig. 4). No malignant cells

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Figure 1. On abdominal radiograph, an arborizing calcification measuring 7×6 cm is located in the left upper abdomen.

Figure 2. On abdominal computed tomography, the mass presenting arborizing calcification is located in the retroperitoneum ventral and inferior to the left kidney.

Figure 3. Gross specimen demonstrates lobulated inhomogeneous tumor with arborizing calcification, 165 g in weight.

Figure 4. (A) Photomicrograph shows scattered lymphoid follicles and abundant small vessels in interfollicular area. (B) Concentric collagenous core with capillaries is seen in center of lymphoid follicle (HE stain, ×30 and ×100).
were seen. The pathologic diagnosis was a Castleman tumor of HV type.

**Discussion**

Retroperitoneal CD represents an uncommon location of a rare disorder. In a review of 315 cases (11), 70% occurred in the mediastinum; 20% in cervical, axillary, shoulder, inguinal, or vulval areas; and 7% (21 cases) in the retroperitoneum. Only 2% of the occurrences (6 cases) involved the pararenal area. Patient age at diagnosis has ranged from 2 to 85 years (median 23) (12); no racial or gender-specific predilection is known (2).

The two histologic types of CD (2) tend to differ clinically. The commoner HV type is usually asymptomatic except for possible compression of adjacent structures. Only 3% of HV type patients present symptoms (13) such as early satiety, abdominal fullness (5), and recurrent urinary tract infections by a curved lateral displacement of the ureter (6). The PC type is symptomatic in approximately 50% of cases, commonly showing anemia, fever, and polyclonal hyperglobulinemia (14).

The HV type is characterized by prominent follicular centers and interfollicular vascular proliferation, while the PC variety consists of normal to large follicles separated by sheets of plasma cells and a somewhat less vascular stroma. The specific etiology of CD remains unclear, although more generally it is thought either to result from a local inflammatory process or to represent a hamartoma of lymphoid tissue (15). Regarding PC type CD, Yoshizaki and Masuda (16) suggested that biochemical products of lymph nodes may cause the clinical abnormalities, and demonstrated that the germinal centers of hyperplastic lymph nodes of patients produce large quantities of interleukin-6. On the other hand, the relation between IL-6 and HV type CD is unclear.

Complete surgical removal is considered the best treatment for localized CD. Even partial excision of unresectable lesions may be useful, since recurrence or progression has not been reported (2, 6).

Preoperative diagnosis of HV type CD is difficult because of its indistinctive clinical features. Radiographically CD most often is indistinguishable from other masses (4–7). The large size of the mass in our case initially made us suspect a sarcoma. Almost one third of CD cases show calcification, and which is commoner in abdominal than in thoracic lesions (13). In our case, abdominal radiographs and CT showed linear radial or stellate calcification similar to that reported as “arborizing calcification” (8–10, 13), a pattern which is emphasized the characteristic for HV type CD and can differentiate from other retroperitoneal masses which may have calcification, such as a teratoma or neurogenic tumors.

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

**References**