Case report

Castleman’s disease in the porta hepatitis

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Abstract. We report the CT, ultrasonographic, and pathologic features of a localized forms of Castleman’s disease in the porta hepatitis due to their unusual location. Our report suggests that the CT and the ultrasonographic features of Castleman’s disease in these locations without calcifications is nonspecific.

Key words: Castleman’s disease – Porta hepatitis – CT

Introduction

Castleman’s disease, an unusual condition of unknown etiology, was first described by Castleman and colleagues in 1954 as a localized mass of mediastinal lymphoid follicles [1, 2]. It has also been called angiolymphoid lymph node hyperplasia, giant lymph node hyperplasia, and lymphoid hamartoma [1, 3]. Castleman’s disease usually occurs in the mediastinum (in 70% of cases), but can occur elsewhere where lymph nodes are normally found [3]. Usually it presents as a large round and single mass [4].

We report the CT, US, and pathologic features of a localized hyaline–vascular type of Castleman’s disease in the porta hepatitis due to their unusual location.

Case report

A 56-year-old woman with ischemic heart disease for 4 years and history of vague abdominal pain for several months was admitted to hospital because of chest pain. Physical examination findings and laboratory tests were normal. No lymphadenopathy or hepatosplenomegaly were noted. Chest X-ray was normal.

The patient underwent abdominal US to examine possible causes of the abdominal pain, which revealed an isolated, hypoechoic, 31 × 26-mm well-circumscribed mass in the porta hepatitis (Fig. 1). Enhanced CT of the abdominal imaging was performed. The scans were obtained with a Toshiba scanner (Toshiba Xvision GX, Tokyo, Japan). The CT confirmed the presence of a mass in the porta hepatic region. The mass, which had central hypodensity without internal calcification, had well-circumscribed outline. After intravenous bolus injection of 50 ml non-ionic contrast medium [Iopromid 300 mg/ml (Ultravist 300), Schering, Berlin, Germany], CT performed in the plane of the lesion demonstrated a pronounced enhancement of the mass, but the density of the hypodense center did not change (Fig. 2).

Fine-needle aspiration with US guidance was performed, but the result was negative. The patient subsequently underwent open biopsy and complete excision of the mass, because the results of the laboratory, fine-needle aspiration, US, and CT did not help to explain the cause of the mass. Surgical exploration revealed an isolated extrahepatic mass located in the porta hepatic space, but no liver abnormality. The mass was completely excised.

Macroscopically, the mass was a well-circumscribed single mass that measured 3.5 × 2.8 × 2 cm. The cut surface was homogeneous and gray-pink, and had nodular patterns in some areas. There was no necrotic area. Histologically, we observed pseudofollicles which were smaller than reactive follicles. The center of pseudofollicles was seen to be composed of a blood vessel with swollen, often proliferated, endothelial cells in concentric arrangement. Closely packed and concentrically arranged small lymphocytes were found around the center (Fig. 3). Some areas had fibrotic bands. The final pathologic diagnosis was Castleman’s disease, localized hyaline vascular variant.

The patient is well 10 months after surgery.
Castleman’s disease has two distinct histologic types; over 90% of cases are of the hyaline vascular type, characterized by small hyalinized follicular center and prominent interfollicular vascular proliferation, whereas the remaining 10% are of the plasma-cell type, characterized by an abundance of plasma cells [2, 3]. Systemic disturbances, including fever, anemia, and hypergammaglobulinemia, are rare in the hyaline vascular type but common in the plasma-cell type [3]. The disease usually occurs in young people. Seventy percent of patients are 30 years of age or younger. The disease is often unifocal [5], but multicentric forms have been described in older men and are more aggressive and sometimes fatal [6]. Our case was a 56-year-old woman.

Castleman’s disease usually occurs in the mediastinum (in 70% of cases) [5]. Outside the thorax, common sites are the neck and retroperitoneum, and rare sites include the pelvis, axilla, muscles – particularly those of the shoulder girdle – the thigh, the cranial cavity, the larynx, and the porta hepatitis. It presents as a large, lobulated, vascular mass with smooth margins [3, 4, 7]. The porta hepatitis separates the quadrate lobe from the caudate lobe and contains the portal vein, the hepatic artery, the hepatic duct, and lymph nodes [7]. The space may be involved by a variety of inflammatory or neoplastic processes. It has been reported that some cases are found incidentally when an X-ray or US is done for other reasons [4], as in our case.

Sonographically, the tumor has been described as a homogeneous hypoechoic mass. Also our case had the same feature. The CT findings of prompt, dense homogeneous enhancement followed by gradual equalization of enhancement have been described [3]. Histologically, Castleman’s disease is most often confused with lymphoma. The CT scans with contrast may be helpful in avoiding this diagnostic pitfall, since lymphoma generally does not enhance on CT scans [1]. Computed tomography scanning also excludes fatty or cystic masses form the differential considerations. The hyperdense appearance on CT would be atypical for most of the extraspinal masses including malignant lesions (e.g., lymphoma, metastasis, cholangiocarcinoma, sarcoma, or malignant fibrous histiocytoma) or benign causes (e.g., sarcoid, localized Castleman’s disease, or infection), except for Castleman’s disease and cholangiocarcinoma, the latter having associated biliary dilatation [3]. Although the central hypodense areas on CT are not diagnostic for Castleman’s disease, it has been reported in the literature [7], as in our case. Calcification is not uncommon in Castleman’s disease and occurs in up to one third of abdominal cases [3]. A radial pattern of calcifications is characteristic in Castleman’s disease on CT. The lesion in our case was uncalcified.

Despite its rarity, abdominal Castleman’s disease can be located anywhere in the abdomen including the porta hepatitis. This rare entity must be considered in a differential diagnosis of pathologic lesions in the porta hepatitis.