Case Report

Retroperitoneal Castleman's Disease

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Keywords

Retroperitoneal tumor · Castleman's disease · Hyaline vascular type

Abstract

Castleman's disease was first reported in 1954 by Castleman et al. and identified as an uncommon lymphoproliferative disorder. In most cases, Castleman's diseases are detected in the chest, head, and neck. A 71-year-old man was referred to our hospital due to a retroperitoneal tumor in the para-aortic area by computed tomography (CT). Positron emission tomography-CT revealed an uptake in this tumor, suggesting malignant diseases. Laparoscopic tumorectomy was performed, and the pathological diagnosis was Castleman's disease, hyaline vascular type. No evidence of recurrence was observed 20 months after surgery. We herein report a rare case of retroperitoneal Castleman's disease.

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Introduction

Castleman's disease was first reported by Castleman et al. in 1954 and identified as an uncommon lymphoproliferative disorder [1]. We herein report a rare case of retroperitoneal Castleman's disease.

Case Presentation

A 71-year-old man was referred to our department for the further examination of his 3-cm retroperitoneal tumor detected by computed tomography (CT) (Fig. 1a). He had no disease history or family history. Laboratory data showed almost all normal findings, including his adrenal hormone level, without the slight elevation of sIL-2R 632 Iu/mL. Positron emission tomography (PET)-CT revealed the tumor to have a maximum standardized uptake value (SUV_{max}) of 3.5 (Fig. 1b), suggesting malignant disease.

Laparoscopic retroperitoneal tumorectomy was performed. During the retroperitoneal approach, the tumor was observed at the lower pole of his left kidney, and no adhesion was observed (Fig. 2). The resected specimen was 35 × 32 × 25 mm and covered with a capsule. The cut surface was yellowish-white (Fig. 3). Histologically, regressed germinal centers and hypervascularity in the interfollicular area were observed (Fig. 4). Based on these findings and the lack of any lymph node enlargement, Castleman's disease, hyaline vascular type, was diagnosed. The patient is free from recurrence 20 months after surgery.

Discussion

Castleman's disease was first reported by Castleman et al. in 1954 and determined to be an uncommon lymphoproliferative disorder [1]. The detailed pathophysiology is still unknown, although IL-6 is suspected to play an important role in proliferative disorders in general [2, 3]. Hamada et al. reported 218 cases of Castleman's disease among Japanese patients. According to that report, the age of onset was typically around 10–40 years old, with no marked gender differences, and the likely occurrence sites were the chest (45.4%), head and neck (24.8%), and retroperitoneal region (11.0%).

Castleman's disease is divided three subtypes: hyaline vascular type (HV), plasma cell type (PC), and a mixture of HV and PC type (Mixed) [4]. HV shows follicular dendritic cell prominence or dysplasia and hypervascularity in the interfollicular regions, and PC shows an increased number of follicles with large, hyperplastic germinal centers. Kawamura et al. reported that HV type was found in 87%, PC type in 8%, and mixed type in 5% of 132 Japanese retroperitoneal Castleman's disease cases.

Castleman's disease is also classified by its distribution as unicentric Castleman's disease (UCD) or multicentric Castleman's disease (MCD). UCD usually shows no phenomenon and no abnormal laboratory findings, so most cases are diagnosed incidentally. In contrast, MCD is associated with the enlargement of multiple lymph nodes, a fever, fatigue, weight loss, among other findings. In addition, laboratory data also show anemia, reduced platelet counts, CRP elevation, hyper γ globulin, and hyper serum IL-6 [5]. These findings are correlated with a pathological type, and 90% of UCD cases are HV type while almost all MCD cases are PC type [6]. The present case showed HV type histologically, and the diagnosis was UCD as no other lesions were observed.

Contrast CT or angiography is reported to show an enhanced tumor with a smooth surface, but PC type is less hypovascular than HV type [7, 8]. Calcification is infrequent and can include punctate, coarse, peripheral, and arborizing patterns. In addition, magnetic resonance imaging of HV type shows intermediate to slightly high signal compared to muscle on T1WI and slightly hyper intense on T2WI [9]. A recent study demonstrated the efficacy of PET-CT for detecting retroperitoneal tumors with a slightly elevated SUV_{max} , although it has difficulty detecting inflammation or malignant diseases. PET imaging of Castleman's disease demon-

strated FDG accumulation in range lower than that seen for low- and intermediate-grade lymphomas [10]. Based on the present findings, a preoperative diagnosis of Castleman's disease was difficult. This case also showed elevated SUV-max 3.5 on PET-CT and was suspicious of being a malignant disease, such as lymphoma.

In all types of Castleman's diseases, surgical resection is recommended and necessary for a differential diagnosis. With the advent of laparoscopic surgery, laparoscopic resection has been selected in recent reports [11]. Castleman's disease HV type was reported to have a favorable outcome after surgical resection, and recurrent cases are rare. However, MCD is reportedly difficult to resect completely, so steroidal treatment and anti-IL-6 receptor antibody tacilizumab are sometimes used, which may prolong the overall survival. This case showed no recurrence 20 months after surgical resection. However, due to the rarity of retroperitoneal cases, careful observation is needed. We herein report a case of retroperitoneal Castleman's disease.

Statement of Ethics

Written informed consent to participate and for publication was obtained from the patient. A copy of the written consent form is available for review from the Editor-in-Chief of this journal.

Disclosure Statement

The authors declare no conflicts of interest.

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Author Contributions

KS, TK drafted the manuscript. RK, JK, SS, RT, HU, KC performed the experiment.

Availability of Data and Material

Due to ethical restrictions, the raw data underlying this paper are available upon request to the corresponding author.

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Fig. 1. Axial CT (a) and PET-CT (b).

Fig. 2. The tumor was adhered strictly.

Fig. 3. Resected specimen.

Fig. 4. Hematoxylin and eosin staining.