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Citation	Surgery Today, 36(7), 647-650 https://doi.org/10.1007/s00595-006-3200-2
Issue Date	2006-07
Doc URL	http://hdl.handle.net/2115/14480
Rights	The original publication is available at www.springerlink.com
Type	article (author version)
File Information	Castlemans_disease.pdf



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Manuscript No. 05-181 **Name: Nagato Sato**

Hyaline Vascular-Type Castleman's Disease in the Hepatoduodenal Ligament: Report of a Case

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Running head: Castleman's Disease

article type: case report

key words: Castleman's disease, hepatoduodenal ligament, preoperative diagnosis

Abstract

Castleman's disease is an uncommon lymphoproliferative disorder, which occurs most commonly in the mediastinum. Sporadic reports of its occurrence in the hepatoduodenal ligament, and other extrathoracic locations have also been documented. Ultimately,

5 Castleman's disease can develop anywhere lymphoid tissue is found, and the preoperative diagnosis is often difficult. We report a case of hyaline vascular-type Castleman's disease in the hepatoduodenal ligament. An asymptomatic 26-year-old woman was admitted to our hospital for further examination of a mass found on ultrasonography. A contrast-enhanced computed tomography (CT) scan confirmed a hypervascular mass in the hepatoduodenal
10 ligament, and angiography showed that the mass was receiving its blood supply from various arteries. These findings suggested a diagnosis of Castleman's disease and we removed the tumor without resecting any other organs. CT and angiography were helpful in establishing a correct preoperative diagnosis, which minimized the magnitude of surgery.

Introduction

Castleman's disease, or angiofollicular lymphoid hyperplasia, is a rare lymphoproliferative disorder of unknown etiology, which was first described in 1956.¹ Histologically, it is classified into two separate subtypes: the hyaline vascular and plasma cell variants, the former being more common and with greater vascularity. It also has two clinical expressions: a localized form, which usually presents as a slow-growing mass and follows a relatively benign clinical course; and a multicentric form, in which tumors are multilocalized, associated with relatively high morbidity and mortality. The lymph node masses most commonly arise in the mediastinum (70%),² but can occur anywhere that lymph nodes are found.³ Extrathoracic locations, such as the peripancreatic site, pelvis, and retroperitoneum, are rare. Establishing a preoperative diagnosis can be difficult. We report a case of hyaline vascular-type Castleman's disease in the hepatoduodenal ligament, for which the magnitude of surgery was minimized by a correct preoperative diagnosis.

Case Report

A 26-year-old woman was referred to our hospital after an ultrasonography during a routine medical check-up showed a mass. She was asymptomatic and her general physical findings were normal, as were her laboratory findings, including a complete blood cell count, serum chemistry, immunoglobulin (Ig)G, IgA, IgE and interleukin (IL)-6. Abdominal ultrasound showed a well-defined, 4 x 4 cm mass, which was elliptic, isoechoic, homogeneous, and adjacent to the liver, gall bladder, biliary duct, duodenum, pancreas, and right kidney. Computed tomography (CT) scans confirmed a well-circumscribed homogeneous soft-tissue dense mass without any calcifications in the hepatoduodenal ligament, by precontrast images. The mass was enhanced by contrast material in the arterial phase (Fig.1). No other enlarged lymph nodes were found. Magnetic resonance imaging (MRI) showed an isolated mass with

smooth margins, which was hypointense on T1-weighted images and isointense on T2-weighted images. On magnetic resonance cholangio-pancreatography (MRCP) the common bile duct was slightly compressed to the right. Endoscopic ultrasound (EUS) showed an isoechoic mass in contact with the gallbladder, bile duct, duodenum, and pancreas, but there was no sign of involvement with them. These imaging findings did not show the origin of the tumor. Angiography showed a hypervascular mass receiving its blood supply from the hepatic, gastroduodenal, and superior mesenteric arteries (Fig.2). The hypervascularity of the mass suggested a diagnosis of Castleman's disease in the hepatoduodenal ligament. We performed a laparotomy, which revealed a hard tumor with abundant arterial feeding from various sources in the hepatoduodenal ligament. We took great care when separating the tumor from the biliary tract because of the dense fibrous adhesion. The tumor was totally removed without resecting any other organs. The total blood loss was 350ml without blood transfusion, and the operation time was 2h 35min. The tumor measured 4.5 x 3.5 x 3.0 cm, and it was removed with the surrounding fibrous capsule. The cut surface was canescent and homogeneous. Histopathologically, the tumor was composed of multiple lymph follicles with concentric layers of mantle zone cells, with an "onion skin" appearance and atrophic germinal centers. The germinal centers had penetrating arterioles with hyalinized vessel walls (Fig.3). These findings were consistent with the localized form of angiofollicular hyperplasia (Castleman's disease) of the hyaline vascular type. The patient had an uneventful postoperative course and was discharged from hospital 10 days after her operation. She has remained free of disease for a follow-up period of 14 months.

Discussion

The hyaline vascular-type Castleman's disease is considered benign, and surgical removal cures the disease.^{2,4,5} However, establishing a preoperative diagnosis is important to avoid

unnecessary organ resection. In our patient, the correct preoperative diagnosis prevented resection of the common bile duct.

The most common site of involvement of hyaline vascular-type Castleman's disease is the mediastinum,² but any lymph node site can be involved.³ To our knowledge, no other case of this disease occurring in the hepatoduodenal ligament has been documented, although there are reports of this disease in the porta hepatis.^{3,6}

It is important to recognize the typical imaging characteristics of this disease.

Ultrasonography generally shows a focal hypoechoic mass with increased through-transmission and small hyperechoic regions,⁷ and CT generally shows a homogeneous soft tissue mass with clear early contrast enhancement, often accompanied by calcification.⁸ The angiographic features are characteristic of a hypervascular mass with numerous enlarged feeding vessels and a homogeneous capillary blush corresponding to the capillary proliferation.⁸ The imaging characteristics found in our patient were typical, except for the absence of calcifications, which enabled us to make a correct preoperative diagnosis, despite the unusual tumor site.

The differential diagnosis of this type of mass includes malignant tumors of the hepatoduodenal ligament, such as liposarcoma, leiomyosarcoma, fibrohistiocytoma, gastrointestinal stromal tumor, and lymphoma. These lesions are usually heterogeneous, with poor contrast enhancement. Lymphoma is often confused with Castleman's disease because it is homogeneous. Thus, the contrasted CT is helpful because a lymphoma is generally not enhanced by contrast material.⁹

The best treatment for Castleman's disease is still unknown.⁵ Surgery has long been considered standard therapy for the localized type, with a variety of case reports and retrospective series reporting excellent rates of cure.^{4,5,10-14} Careful follow-up with regular confirmation that the tumor is not growing might be an alternative to a hurried surgical

intervention, as long as the characteristic findings of diagnostic imaging remain unchanged, although there have been no reports of long-term follow-up without surgery. However, with follow-up alone, there may be a risk that tumor growth could evoke clinical symptoms caused by a mass effect, which could make the surgical magnitude greater, necessitating other organ resection or making the tumor unresectable. Moreover, cases of malignancy developing in the localized form of hyaline vascular-type Castleman's disease have been reported.¹⁵ Thus, complete resection soon after the initial diagnosis is made is likely to afford the best chance for cure.

The most difficult part of the resection is the fibrous adhesion and hypervascularity of the tumor. The adhesion of the tumor to the surrounding tissue, despite its encapsulation, is a feature associated with Castleman's disease.¹⁶ Moreover, the adhesion and hypervascularity may precipitate massive bleeding on excision. Therefore, transfusions should be prepared for the potential loss of blood. Although our patient had tight adhesions between the tumor and the surrounding tissue, including the biliary tract, with much bleeding from the vessels near the mass, the tumor was completely removed while preserving the biliary tract, and the patient did not require a blood transfusion. Whereas angiographic preoperative embolization has been reported to make surgical excision easier by minimizing intraoperative bleeding,¹⁷ surgery designed with great care for hypervascularity is considered important to prevent massive bleeding.

In conclusion, Castleman's disease can be located anywhere that lymphoid tissue is found. This rare entity should be included in the differential diagnosis of hypervascular, well-demarcated soft tissue masses.

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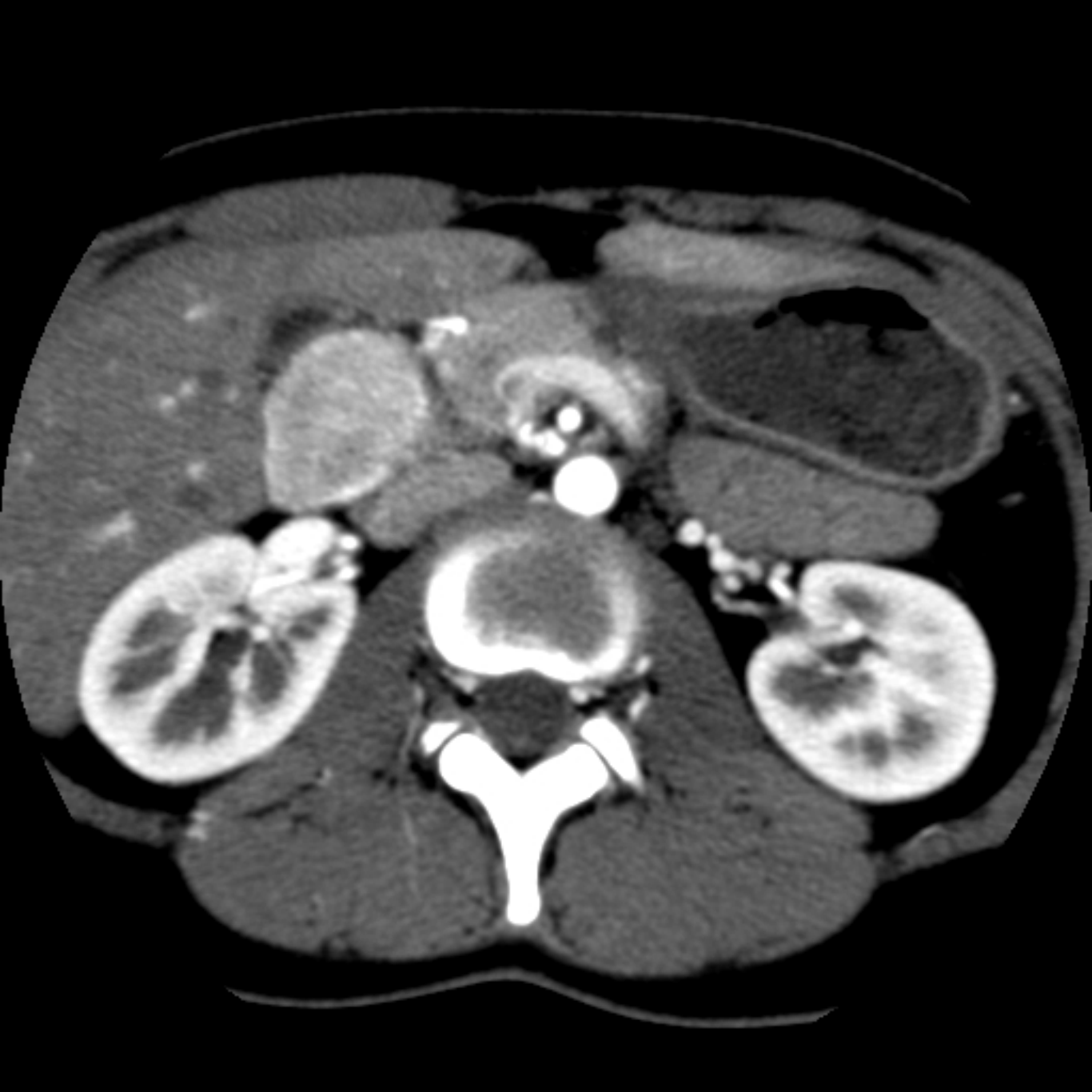
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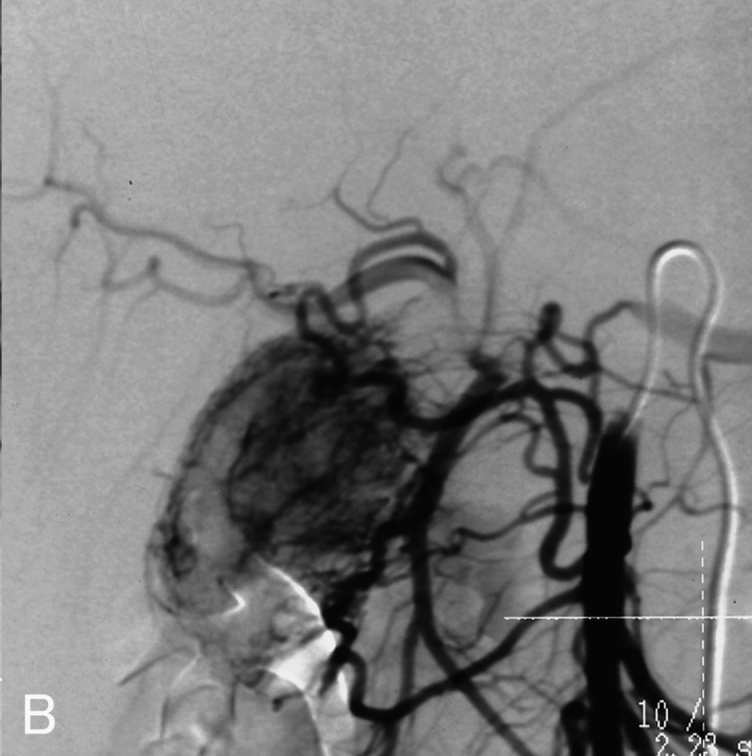
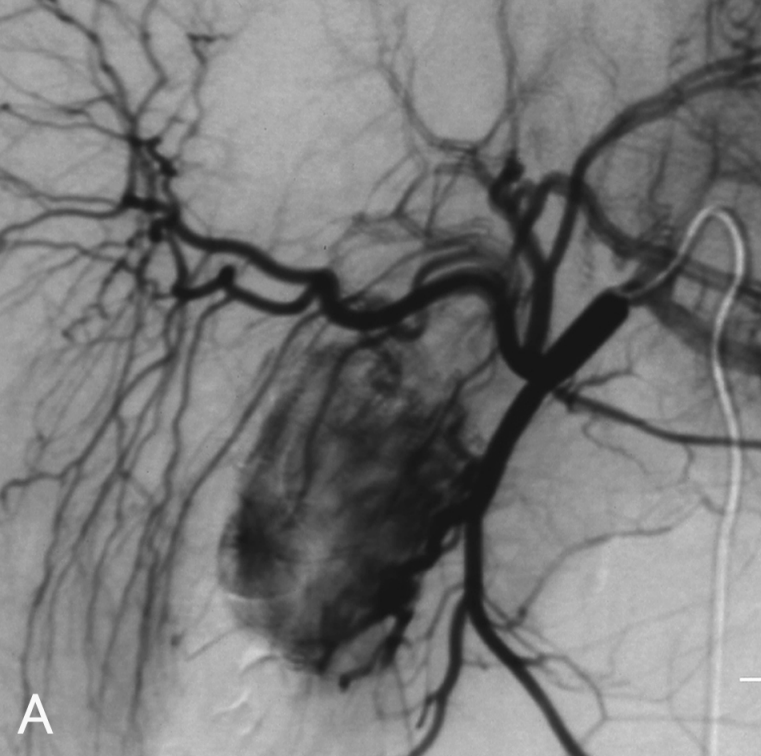
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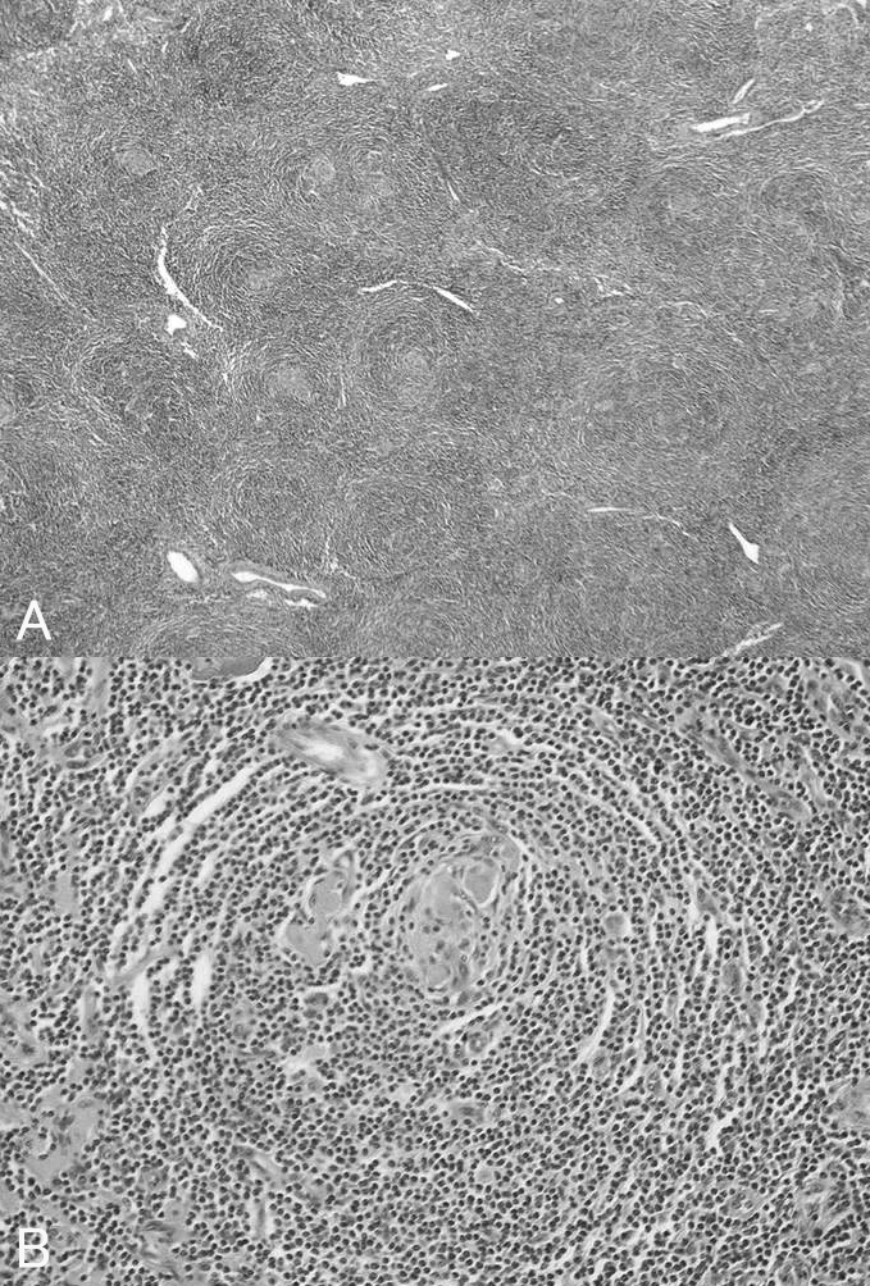
Fig.1: Computed tomography images confirmed a well-circumscribed, homogeneous, soft-tissue mass in the hepatoduodenal ligament. The mass was dense and remarkably enhanced by contrast material in the arterial phase.

5 Fig.2: Selective angiography of the celiac (**A**) and superior mesenteric arteries (**B**) showed a hypervascular mass receiving its blood supply from the hepatic, gastroduodenal, and superior mesenteric arteries.

Fig.3: The tumor was composed of multiple lymph follicles with concentric layers of mantle zone cells, giving an “onion skin” appearance, and atrophic germinal centers. The germinal
10 centers had penetrating arterioles with hyalinized vessel walls. (**A** H&E, ×100 **B** H&E, ×400)







A

B