Primary Castleman's Disease in the Hepatis Caudate Lobe: A Case Report

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Case report

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Abstract

Background: Castleman's disease (CD) is an uncommon type of benign proliferation of the lymphoid tissue.The occurrence of CD in the liver is rare. Radiological diagnosis of hepatic CD by computed tomography (CT) and magnetic resonance imaging (MRI) remains difficult. It is difficult to distinguish between CD of the liver and hypervascular liver tumors such as focal nodular hyperplasia (FNH) and hepatocellular adenoma before surgery.

Case presentation: We report a unique case of CD in the hepatis cauda lobe in a 26-year-old young woman after a health checkup,abdominal ultrasonography, CT and MRI results showed a 5 cm×3 cm sized hypervascular mass. The patient was diagnosed as CD by histologic examination. The liver mass showed the imaging feature of central low density or signal enhancement in the arterial phase on contrast-enhanced CT and MRI, the tortuous access vessel could be seen in the lesion. The tumor became homogeneous isodense or isointense to the surrounding liver tissues during the portal phase. Dffusion restriction demonstrated on MRI. All these imaging findings, which might be useful clues for identifying CD in the liver.The patient underwent complete resection of the tumor. The patient was followed-up without any further treatment and the disease has shown no signs of recurrence.

Conclusions: Primarily hepatic CD without lymph node hyperplasia or clinical symptoms is extraordinarily rare and different from other hypervascular liver tumors. Therefore, it is extremely important to improve the awareness of this diagnosis. Attention should be paid to the difference hepatic CD between FNH, hepatocellular adenoma, lymphoma and so on. To avoid misdiagnosis with unnecessary surgical excision, distinguishing these diseases is crucial.

Background

Castleman’s disease (CD, also known as vascular follicular lymph node hyperplasia, angiofollicular hyperplasia and giant lymph node hyperplasia) is a rare type of benign proliferation of the lymphoid tissue that was first reported in 1954[1, 2]. It has been classified into five distinct subtypes on the basis of histologic forms: hyaline vascular CD, plasma cell CD, mixed-type CD, human herpes virus 8-associated CD and multicentric not otherwise specified CD[3]. The hyaline vascular type accounts for approximately 90% of all patients with CD[1].Clinically, it is classified into unicentric and multicentric types[4]. It can appear in any part of the body, but usually occurs in the neck, mediastinum, and lungs[5]. The cases of CD in the liver are extremely rare, due to lack of specific clinical symptoms, laboratory examination results and radiological findings, radiological findings, easily misdiagnosed before the operation, often requires clear diagnosis by histopathology after resection. Histopathology is the gold standard of diagnosis, which characteristically manifests as the enlargement of numerous lymphoid follicles with expanded mantle zones and consequent atrophic centers and follicle centers penetrated by increased vessels with fibrous hyalinization[6].

Case Report

In a 26-year-old female patient, a mass was found in the hepatis cauda lobe with size of about 5 cm x 3 cm detected by ultrasound after a periodic health checkup in Sir Run RunShaw Hospital,Zhejiang University School of Medicine. She had a rectal adenoma resection history ten years ago, but without any clinical discomfort, such as nausea, vomiting, abdominal pain, abdominal distention, fever, chills. The abdomen was flat, soft, and not tender. No noteworthy neurological abnormalities were found. She had no history of family genetic disease, occupational disease or drug allergy. Besides, tumor markers including CA125, CA199 and AFP, CEA wereall normal, and laboratory examinations including blood routine examination, function of liver and kidney were all within normal ranges. By abdominal ultrasonography, a solid, well-circumscribed hypoechoic mass approximately 5 cm × 3 cm in size was noted in the hepatic S1. The mass located slightly highlight the outline of the capsule, form the edge of the rules. Abundant blood flow signals were present inside the lesion (figure1). A plain CT scan of the abdomen revealed a sharply demarcated low absorption area approximately 5 cm × 3 cm in size visible in the S1 without haemorrhage and necrosis (figure 2A). There were no obvious swollen lymph nodes in the porta hepatic and retroperitoneal areas. On dynamic CT scan, a thin membrane-like contrast enhancement was noted in the periphery during the arterial phase although slightly low density was seen inside the mass (figure 2B). The tortuous access vessel could be seen in the lesion. The tumor was homogeneous isodense to the surrounding liver tissues during the portal phase (figure 2C). On abdominal magnetic resonance imaging (MRI), a mass approximately 5 cm × 3 cm in size was visible in the hepatic S1 as a low signal intensity area on nonenhanced T1-weighted images (figure 3A) and as a slightly high

signal intensity area on T2-weighted images (figure 3B). This tumor had restricted on diffusion weighted imaging (figure 3C). The features of dynamic enhanced MRI scan for mass were similar to that of dynamic computed tomography scan (figure 3D-F).

Finding with resected specimens

Laparoscopic resection of hepatoduodenal ligament mass was performed: a tumorous lesion approximately 5. 0 cm in diameter above the head of the pancreas was observed beside the hepatoduodenal ligament, in the frontal hepatic cauda lobe, with complete capsule and clearly distinguishable from surrounding tissues was noted. Yellowish-white necrosis areas were found in the tumor tissue after incision. No significant swelling of lymph nodes was observed.The histopathology of intraoperative freezing sections indicated that "lymphoma (hepatic caudate lobe mass) cannot be excluded, and further diagnosis should be performed routinely".

Histopathological findings

Pathological hematoxylin-eosin(H&E) was performed for microscopic examination. The lymphoid follicles were increased, with an expanded mantle zone of small lymphocytes and atrophic germinal centers, resulting in an "onion skin-like" structure (Fig.4). Molecular pathology BCR: IgH gene rearrangement, IgH Fr2A+Fr2B -, IgH Fr3A -, gK gene rearrangement, IgK-. Immunohistochemical results: interfollicular zone cells CD3(+) (Fig. 5A), follicular germinal center cells CD20(+) (Fig. 5B), follicular dendritic cells CD21(+) (Fig. 5C), ki67 (the germinal center is highly proliferative) (Fig. 5D), CD30(-), BCL-6(focal+), MUM1 (focal+), kappa (-), lambda (-). On the basis of these findings, the patient was diagnosed as CD (hyaline vascular type) in the hepatis caudate lobe, a very rare condition. This case was treated using surgical resection. The patient did not receive postoperative adjuvant radiotherapy and chemotherapy. At the present time, the patient is followed-up without any further treatment and the disease has shown no signs of recurrence.

Discussion

Castleman's disease (CD) was first described by Benjamin Castleman in 1954. It is characterized by hyperplasia of lymphoid tissue. It had a good prognosis but without an identified cause. The sites frequently affected by this disease were reported to be the neck, mediastinum and hilum of lung. Primary hepatic CD is very rare with less than 20 cases reported worldwide by Hisaaki et al[7-10]. The development of this disease inside the hepatis, as in the present case, may be viewed as very rare[11]. According to the case reports for localized CD in the hepatic(Table 1), most cases had no obvious clinical symptoms , most of them were hyaline vascular type and women over 40Y.

Clinical and pathological features of CD

Histological evaluation plays a vital role in the exact diagnosis of CD. The histopathological features consistent with CD include abnormal regressed or hyperplastic germinal centers, follicular dendritic cells prominence, hypervascularisation, expanded mantle zones, and interfollicular plasmacytosis . Clinically, it is classified into unicentric and multicentric types. Cases of this disease with a single lesion usually have good prognosis, while cases with multiple lesions tend to have poor prognosis. Most cases with the single lesion are pathologically rated as hyaline vascular type, while those with multiple lesions are considered as the plasma cell type. The hyaline vascular type occurs most frequently as a focal lesion that is not associated with obvious clinical symptoms and is often incidentally found during routine physical examination, while the plasma cell type is often accompanied by systemic symptoms including fatigue, fever, night sweats, weight loss, joint pain and hepatosplenomegaly. This case we described was without any discomfort, however on the basis of these pathological findings, the diagnosis of hyaline vascular type was supported. It was also consistent with previous literature reports[12-16].

Imaging features of CD

No finding from diagnostic imaging specific to hepatic CD has been known. Imaging manifestations of the disease are very difficult to distinguish it from other diseases, including FNH, hepatocellular adenoma. However, ultrasound, CT and MRI can still provide reference values for the diagnosis of hepatic CD. When examined by ultrasonography, the affected area is often depicted as a well-

demarcated hypoechoic mass with a homogeneous inside. In addition, ultrasound also has some advantages in estimating the location of the mass and its adjacent feeding vessels. On CT scan, it often presents as a single, well defined soft tissue lesion, with rare cystic degeneration and focal necrosis. The plain CT scan images of this disease show low homogeneous density lesions. When examined by MRI, this disease is often seen as a low signal intensity area on T1-weighted images and a high signal intensity area on T2-weighted images. This tumor had restricted diffusion on diffusion weighted imaging. Contrast-enhanced CT/MRI of the hyaline vascular type lesions display obvious enhancement, the mean CT value was increased from 47HU to 126HU. Our review of the relevant literature also demonstrated hypervascular mass in almost all cases(Table1). During the arterial phase, the tortuous access vessel could be seen in the lesion. One case showed a 3.3 cm × 2.4 cm sized well-enhanced mass with central vessels(Table 1). The lesion exhibited inhomogeneous enhancement with low density or sign area in the center on enhanced CT/MRI images during the arterial phase. The tumor was homogeneous isodense/isointense to the surrounding liver tissue during the portal phase, the mean CT value was 127HU, and showed persistent enhancement during the delayed phase. Three cases showed over 3. 0 cm sized well-enhanced mass with central low density. Another revealed a 1. 5 cm × 1. 0 cm sized low-attenuated mass. This finding may be associated with the small arterial lumen and the subsequent reduction in blood velocity. In addition, some tumors accompanied calcifications, a 43 year-old female patient had a 8. 0 cm × 10. 0 cm × 13. 0 cm size mass with bulky, dendritic calcifications. Another a 48 year-old female patient had a 5.0 cm diameter mass with small calcifications. There was no obvious calcification in this case. The liver mass showed the imaging feature of central low density or signal enhancement and in the arterial phase on contrast-enhanced CT and MRI, the tortuous access vessel could be seen in the lesion. The tumor became homogeneous isodense to the surrounding liver tissues during the portal phase. The mass showed high signal intensity on diffusion-weighted imaging, which might contribute to the differential diagnosis of hepatic CD from other benign hypervascular tumors.However, pathological examination is required for a definitive diagnosis.

Treatment methods

The management of the unicentric form is resection[17], so the patient underwent complete resection of the tumor and did not undergo radiochemotherapy after surgery. As in this case, the patient was followed-up without any further treatment and the disease has shown no signs of recurrence.

Conclusion

Since hepatic Castleman's disease has been reported rarely worldwide, the lack of understanding of the disease, coupled with the absence of specific signs in the imaging manifestations, makes operative diagnosis extremely difficult. Through this case report, the understanding of the disease can be improved, which will provide a significant reference for future cases. Our case suggests that CD should be considered as a rare differential diagnosis in patients with a hepatic hypervascular mass.

Declarations

Authors' contributions

Jian Wang contributed to this manuscript: literature research, case management, case collection, Bailing Dai contributed to this manuscript literature research, and writing of the manuscript. Fang Yang contributed to this manuscript: pathological images collection procedure. Wenming Zhang contributed to this manuscript: radiological images collection procedure. Jian Wang and Xiandi Zhu contributed to this manuscript: critical revision of the manuscript. All authors read and approved the final manuscript.

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Competing interests

The authors declare that they have no competing interests.

Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable requests.

Consent for publication

The authors thank the patient for informed consent to this publication and the related images.

Ethics approval and consent to participate

Not applicable. Due to the treatment of a single patient with approved surgical treatment , no ethics approval was obtained. The authors thank the patient for informed consent to this publication and the related images.

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Tables

Table.1

Cases of Localized Castleman's Disease in the hepatic

	Clinical presentation	Sex/age	Size（cm）	Calcifications	Enhancement	Central low density	vessel	DWI	Pathology
1	no	54Y/F	2.2×1.87	no	hypervascular	no	no	high	HV
2	abdominal pain	40Y/F	3.0×2.0×2.0	no	hypervascular	no	no	/	HV
3	no	57Y/F	3.3×2.4	no	hypervascular	yes	yes	high	HV
4	abdominal pain	48Y/F	5.0×5.0	small	hypervascular	no	no	/	HV
5	no	28Y/M	3.0×3.0	no	hypervascular	yes	no	/	HV
6	nausea, vomiting,and abdominal pain	43Y/F	8.0×10.0×13.0	dendritic	hypervascular	no	no	/	HV variant+PC variant
7	abdominal pain	56Y/F	3.1×2.6	no	hypervascular	yes	no	/	HV variant
8	No	69Y/F	1.5×1.0	no	hypervascular	yes	no	high	HV
9	No	64Y/F	1.4×1.4	no	hypervascular	no	no	high	HV variant
our case	No	26/F	5.0×3.0	no	hypervascular	yes	yes	high	HV
F, female; M, male; HV, hyaline-vascular; PC, plasma-cell									

Figures

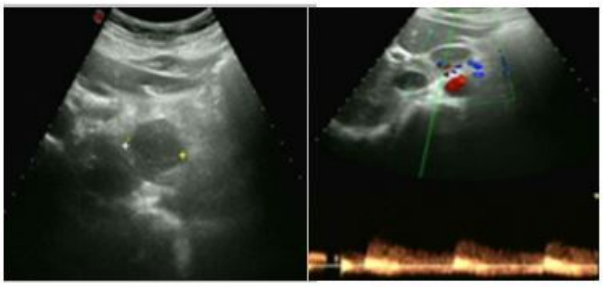

Figure 1

A solid, well-circumscribed hypoechoic mass approximately 5cm × 3 cm in size was noted in the hepatic S1. The mass formed the edge of the rules. Abundant blood flow signals were presented inside the lesion.

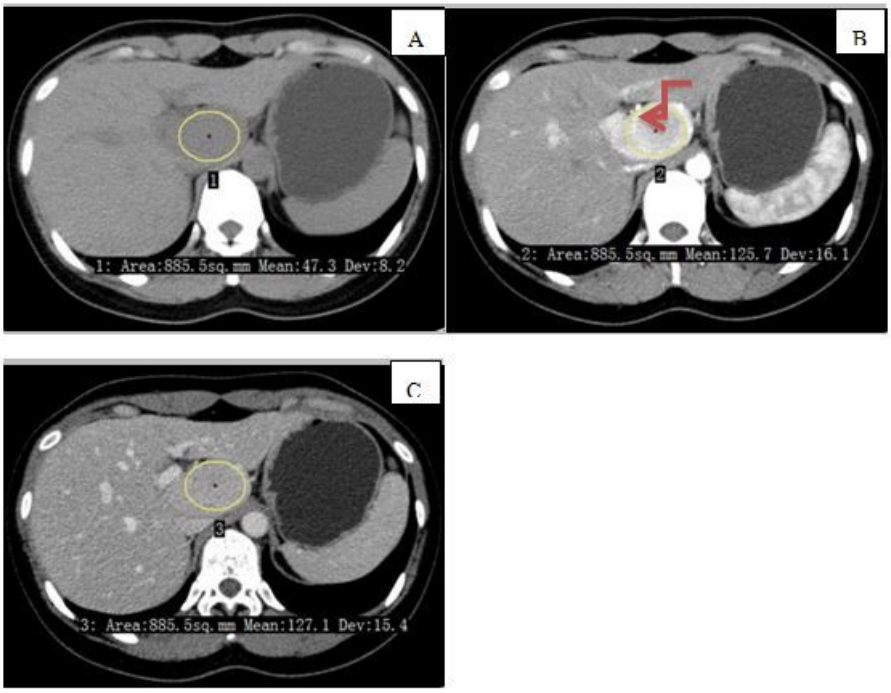

Figure 2

A: a sharplydemarcated low density area approximately 5cm × 3 cm in size was visible in the S1, the mean CT value was 47HU. B:In arterial phase, evident contrast enhancement was noted in the periph¬ery but not inside the mass, the mean CT value was 126HU.The tortuous access vessel(arrow)might be seen indistinctly in the lesion. C: The tumor heterogeneous iso¬dense to the surrounding liver during the portal phase compared with normal hepatic prenchyma was noted. The mean CT value was 127HU.

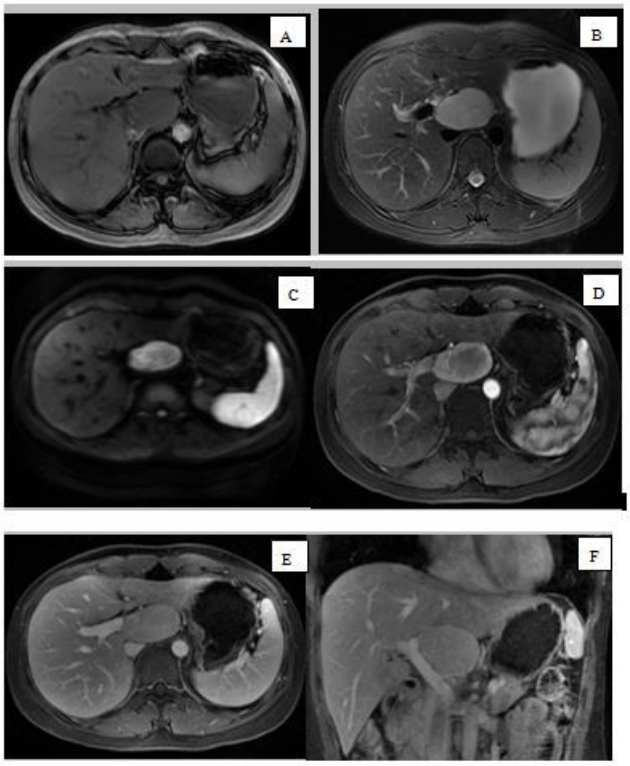

Figure 3

A mass approximately 5cm × 3 cm in size was visible in the hepatic S1. A: Low signal inten¬sity on T1WI. B: High signal intensity on T2WI. C:Diffusion-weighted imaging (DWI) was limited. D:In arterial phase, heterogeneous reinforcement for the mass. E:In portal phase, the tumor was ho¬mogeneous isodense compared with normal hepatic parenchyma. F:Coronal scan for the mass.

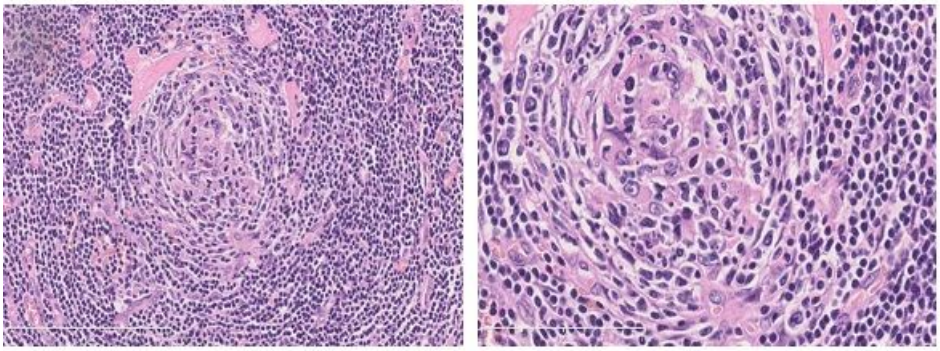

Figure 4

Histopathological findings. A: The lymphoid follicles were increased, with an expanded mantle zone of small lymphocytes and atrophic germinal centers, resulting in an "onion skin-like"structure(HE staining, X200) B: The strip like area was composed mainly of lymphocytes(HE staining, X400).

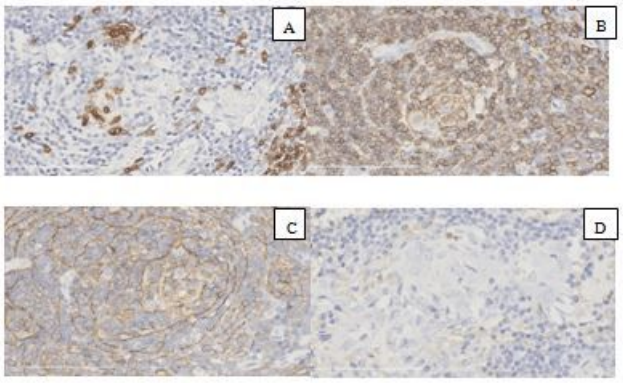

Figure 5

(The hepatic mass immunohistochemical staining, X400) A: interfollicular zone cells CD3(+), B: follicular germinal cen¬ter cells CD20(+), C: follicular dendritic cellsCD21(+) D: ki67(the germinal center is highly proliferative).