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Perigastric Rare Disease with Lymphoproliferative: Hyaline Vascular Type Castleman Case Report

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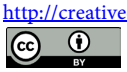


Abstract

Castleman's disease (CH) is a rare chronic lymphoproliferative disease whose etiology is often unclear and whose definitive diagnosis can be made immunopathologically. CH was first described by Castleman *et al.* in 1956 as a non-lymphoproliferative disease. It can be easily misdiagnosed as a gastrointestinal stromal tumor (GIST), which is submucosal in the abdomen, especially when found in the perigastric area. Although patients are often asymptomatic, the diagnosis is usually made by abdominal ultrasonography (USG) and computer tomography (CT) performed for routine screening. Preoperative diagnosis is very difficult due to nonspecific imaging findings and rarity. In our patient who was diagnosed with CH after USG, CT, EUS (endoscopic ultrasonography) and laparoscopic excision and immunopathological analysis, we see the contribution of EUS to our diagnosis in the preoperative period in addition to routine diagnostic tests.

Subject Areas

Medicine & Healthcare

Keywords

Castleman's Disease, Hyaline Vascular Variant, Gastric Lymph Node, Case Report

1. Introduction

Castleman's Disease (CH) is a rare disease characterized by lymph node hyperplasia, although its pathogenesis has not been clearly clarified since it was de-

scribed by Castleman in 1956 [1]. CH is divided into 3 types according to histopathologic characteristics. Among these types which are unicentric, multicentric and HHV-8 associated, we will present our patient with hyaline-vascular type which is a sub-variant of the unicentric type. Although hyaline vascular type is 1.5 times more common in women than in men, it is most common in the 3rd-4th decade. It is most commonly seen in the mediastinum (30%), followed by the neck and abdomen (20%). It is usually asymptomatic and rarely associated with B symptoms [2]. Also a group of HIV-negative and HHV-8-negative CD patients whose disease we propose referring to as idiopathic CD. The etiology of iCD proinflammatory hypercytokinemia is not known and may be viral, inflammatory, or neoplastic [3]. The optimal treatment is curative surgical resection. We present a 40-year-old female patient who was cured after operation.

2. Case Presentation

A 40-year-old woman complained of swelling and pain in the epigastrium that started in 2020. The patient, who had multiple doctor visits and received many medical treatments in this subject, underwent an endoscopy in 2021 after the treatment did not work and the result was normal. Upon this result, the patient even sought help from a psychiatric outpatient clinic. The patient had no previous gastrointestinal history and no special medical, familial, genetic or psychosocial history. Since the patient complained of dyspepsia, no mass was detected on medical examination and there was no abnormality on chest X-ray, a diagnostic USG was recommended. After USG performed on an empty stomach revealed hydronephrosis in the right kidney, MRI and CT were ordered for further nephrological analysis. CT and MRI of the abdomen revealed a solid lesion adjacent to the stomach and pancreas. Tomography was performed for a clearer understanding of the relationship between the lesion and the stomach wall.

On CT images, diffuse concentric wall thickness increases were observed in the distal part of duodenum part 3 and part 4. There was a well-demarcated homogeneous soft tissue mass measuring approximately 40 × 20 × 23 mm at the level of the small curvature of the stomach. Pancreas size and parenchyma were normal. When the solid lesion defined at the level of the small curvature was observed as isodense with the spleen and evaluated together with dynamic MRI findings, it was primarily interpreted in favor of the accessory spleen. Follow-up of the case was recommended, but since intense contrast intensity was observed in the arterial phase and equal contrast intensity was observed in the venous and delayed phases, it was decided to perform EUS imaging for this lesion. EUS imaging showed a lesion bordering the stomach, liver and pancreas, approximately 26.5 × 20.5 mm in size, with a smooth contour, hypoechoic structure and vascular flow on Doppler imaging, and EUS-FNA was performed on this lesion with 22 G needle capillarization method.

As a result, immunohistochemical examinations of the sent tissue samples showed T and B lymphocytes at approximately similar rates, and the findings

described did not lead to a specific diagnosis. Clinical evaluation of the case and excisional biopsy were recommended if necessary. Since the lesion was in close proximity to the stomach and pancreas, excision was decided considering the possibility of gastrointestinal stromal tumor.EUS images are shown in Figure 1 and Figure 2.

The patient was prepared for laparoscopic surgery and taken into operation. Intraoperatively, a nodule attached to the small curvature of the stomach was found; dilated and tortuous vessels were seen on and around the surface (shown in Figure 3). The lesion was removed without any problems (shown in Figure 4), and the final paraffin pathology result was a CD of hyaline-vascular type, hematoxylin-eosin staining showed obvious vascular proliferation and hyalinization in the abnormal germinal center, with tight concentric layer of lymphocytes around the follicle, with accumulation of CD21+ (FDC), consistent with the hyaline-vascular type of CD (shown in Figure 5). At follow-up at 10 months after laparoscopic excision, the patient recovered very well with no evidence of recurrence and no problems at any point from the time of diagnosis to the operation.

3. Discussion

CH, which is a rare disease characterized by lymph node hyperplasia but whose pathogenesis has not been fully clarified and is thought to be due to

Figure 1. Endoscopic ultrasonography images.

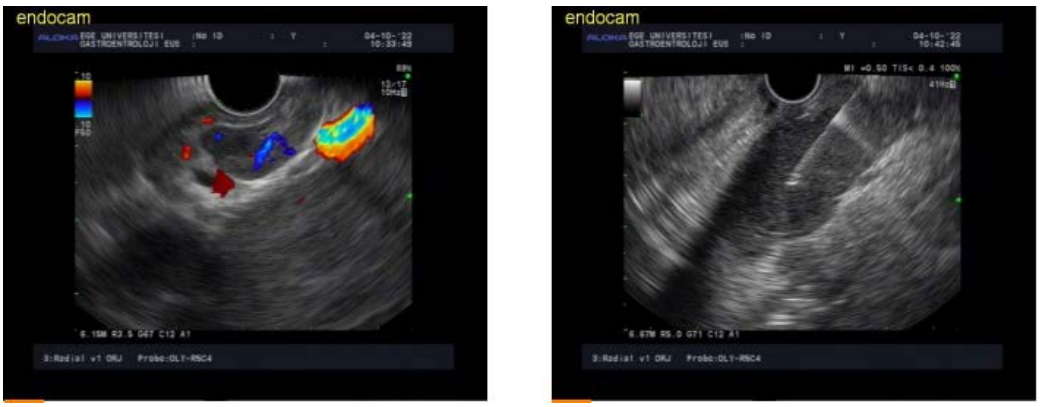
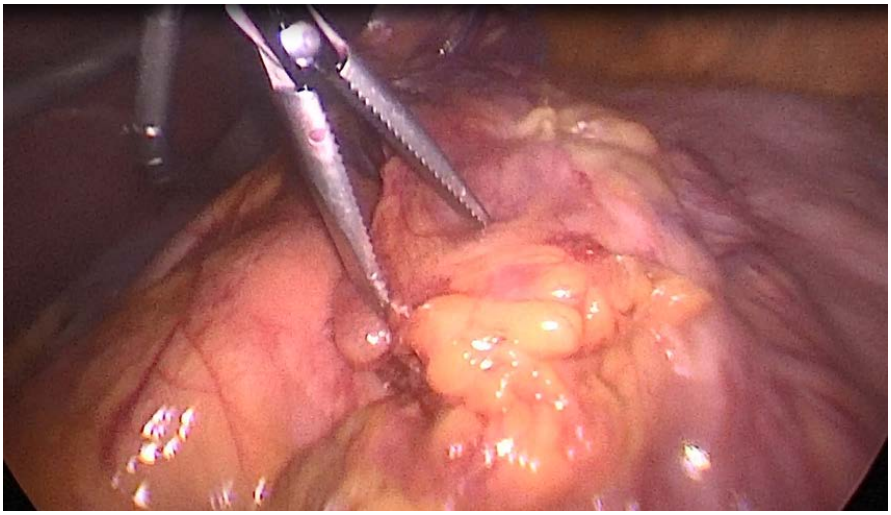
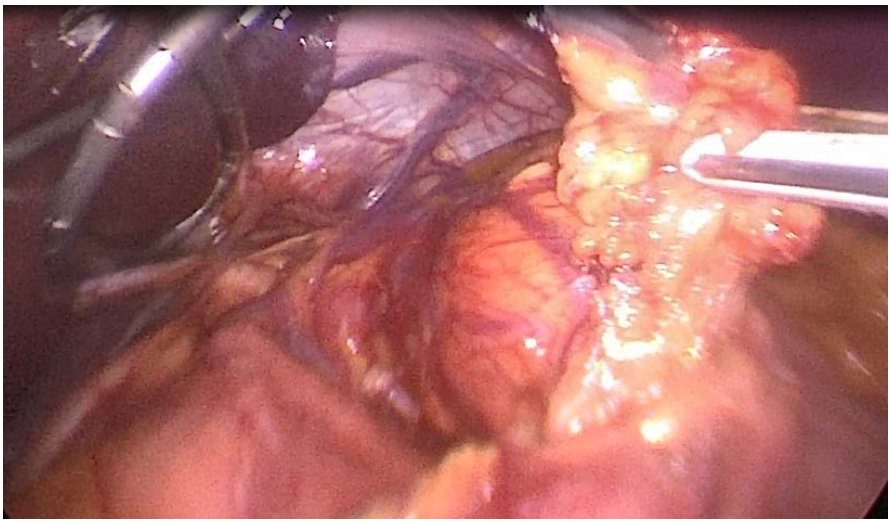


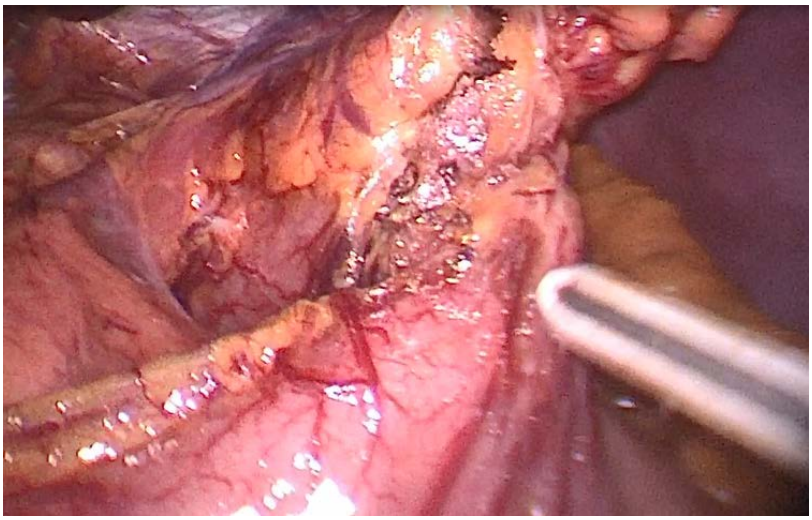
Figure 2. Eus-doppler images.



(a)



(b)



(c)

Figure 3. İntraoperative images.

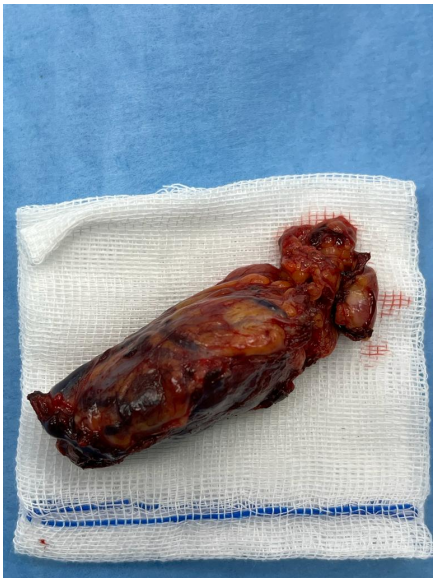


Figure 4. Excision material.

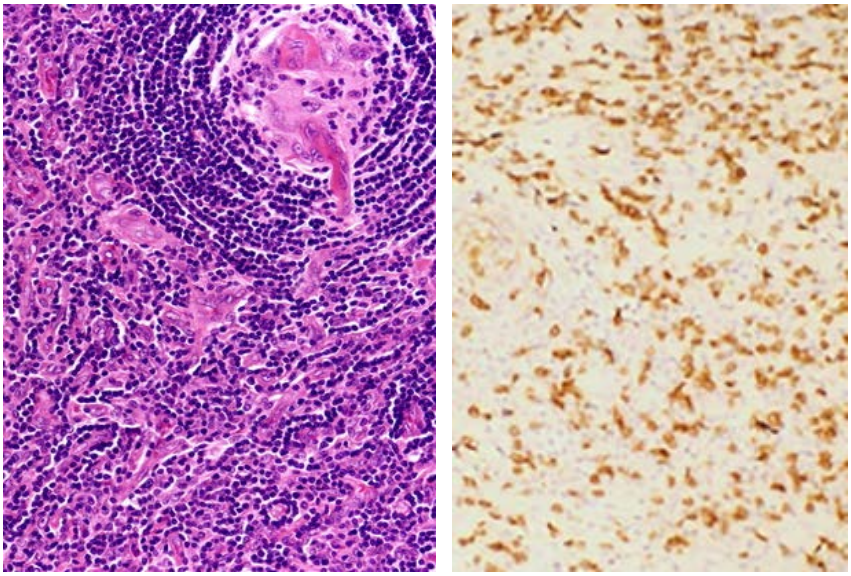


Figure 5. Histopathological and immunohistochemical images

chronic inflammatory stimulation, viral infection and abnormal cytokine release, is usually characterized by painless lymphadenopathy without systemic symptoms [3]. When the subtypes of this rare disease are examined, the most common type is the hyaline-vascular type. When examined pathologically, we see that in this type, lymphatic follicles proliferate in a disorganized manner, there are no germinal centers in these lymphatic follicles and they appear as small transparent vascular follicles. Although not typical in immunohistochemistry, CD34 (marking vascular hyperplasia) and CD21 (marking follicular dendrites) are more commonly visualized.

We have shown that EUS can be used in the preoperative diagnosis of this disease, which is rarely reported in the literature, in addition to USG and CT. As

in our patient, we can more easily examine the structure of the lesion on EUS. The fact that the lesion is solid, hypoechoic, homogeneous, well-circumscribed, with prominent vascularity and increased elasticity on EUS shifts our diagnosis more towards unicentric CH. Another advantage of EUS that we used in our patient is fine needle biopsy (FNB) in the same session, but as in all lymphomas, FNB is insufficient for subtype classification in our case and does not eliminate the need for surgery. We aimed to show once again that EUS can be used in the differential diagnosis of these lesions, which are generally misdiagnosed as GIST on USG and CT. However, despite its advantages, the fact that it is insufficient in terms of biopsy sampling is obvious and our study supports this [4].

The importance of CT in the preoperative evaluation of Castleman's disease is indisputable. In addition to being diagnostic, CT is important in evaluating the surgical resectability of the lesion in the preoperative period. CH is generally observed with a smooth surface and homogeneously distributed density on tomography [5]. This is supported by our study.

4. Results

CH originating from the abdomen appears as a lesion due to lymphoproliferation on imaging methods. The differential diagnosis includes gastrointestinal stromal tumor (GIST), gastric schwannoma, gastric leiomyoma, and lymphadenopathy (LAP) due to other etiological causes (desmoid, inflammatory, metastatic). GIST has unclear borders with the submucosal origin of the stomach, but this is difficult to differentiate on USG due to the homogeneous growth pattern and delayed washout of contrast medium in small tumors. In this case, adding CT and EUS to imaging will help in both identification and preoperative surgical planning [6]. We have seen the diagnostic help of EUS in our case.

"Take home" clinical message the only curative treatment option for unicentric hyaline-vascular variant CH is surgery with complete resection. In cases that cannot be completely resected, radiotherapy is a good treatment option with a 50% complete response success rate. In patients who do not respond to treatment, systemic therapies applied for multicentric CH can be adjusted. It is important to follow up with the patients at the 1st postoperative month and then annually for at least 5 years [2].

Ethics

Informed Consent: Informed consent was obtained from the participant before inclusion in the case report.

Peer-Review

Externally peer-reviewed.

Financial Disclosure

The authors declared that this study received no financial support.

Conflicts of Interest

The authors declare no conflicts of interest.

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