LESS

Case Report

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Totally laparoscopic approach for a single mesenteric localization of Castelman's disease: state of the art of laparoscopic surgery

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ABSTRACT

Castleman's disease is a rare proliferative disorder of benign nature characterized by hyperplasia of the lymphoid follicles. The abdomen is rarely affected by this pathology. The difficult preoperative diagnosis has recently led laparoscopic surgery to become an option for diagnosis and treatment of the unicentric form of the disease. Herein we present a literature review reporting our experience representing, according to our knowledge, one of the first cases of totally laparoscopic resection of single abdominal localization of Castelman's disease.

Keywords: Castleman's disease; laparoscopy; surgery.

Introduction

Castleman's Disease (CD) represents a distinct clinic-pathological entity, first described by Dr. Benjamin Castleman in 1956,[1] which still remains a rare and poorly understood disease characterized by massive growth of lymphoid tissue. A variety of terms have been used to describe this disorder, including giant lymph node hyperplasia, lymph node hamartoma, follicular lymphoreticuloma, benign giant lymphoma, angiomatous lymphoid hamartoma, and angiofollicular mediastinal lymph node hyperplasia.[2] The most frequent location of the CD is the chest (70%), although this pathology can theoretically affect any nodal station. The abdomen is rarely affected by this pathology; abdominal localizations reported in the literature are: mesentery, retroperitoneum, pancreas, pelvis and rectum.[3]

We conducted a review of the English literature articles of CD treated with laparoscopic surgery using PUBMED and Google SCHOLAR databases from the first publications until January 2018.

The words used for our research were: “Castleman’s Disease” and “laparoscopy”. We found 14 cases of Castleman’s Disease treated with video assisted approach. It is exceptionally uncommon for Castleman’s disease to present in the mesentery and only 54 cases except ours

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have ever been described in the literature, with 52 of them that underwent a laparotomic excision, and two cases with a laparoscopic-assisted approach.[4]

There are three pathologic variants of Castleman's Disease: hyaline vascular CD, plasma cell CD, and mixed type of CD, which is characterized by the presence of both hyaline vascular and plasma cell CD types. Plasmablastic variant of CD, which is considered as a sub-variant of plasma cell type, occurs predominantly in immunosuppressed patients and human immunodeficiency virus (HIV)-positive patients.[5] Clinic-pathologically, we can find 3 types of variants: (1) Localized CD of the HV type, presenting as an asymptomatic and slow-growing mass, (2) Localized CD of the PC type, featuring systemic manifestations of inflammation, and (3) B-cell hyperreactivity. The third group is also referred to as Multicentric Castleman's Disease (MCD) and is characterized by systemic lymphadenopathy, frequent multi-organ involvement, and more aggressive behavior.[6] The diagnosis is difficult and the diagnostic certainty is obtained only by histological examination. Furthermore, there is no gold- standard treatment for this disease.[3]

Surgery is the optimal therapeutic approach only in the localized form, while for unresectable or disseminated disease, partial surgical resection, steroid, chemotherapy and radiotherapy have been employed with some measurable success.[7]

This report describes a case of a mesenterial localization of the hyaline vascular CD in the localized form.

Case Report

A.A., a 28 years old man with a history of Bechet Desease HLA B51 positive, spastic-ataxic paraparesis, treated with immunosuppressive therapy for several years, was submitted to abdominal ultrasound during investigations for a fever non responsive to drugs. The abdominal ultrasound highlighted the presence of a 4 cm nodule in the right iliac fossa without abdominal pain.

The Patient underwent a CT abdominal scan that shows a 4 cm inhomogeneus nodule near the duodenum and multiples nodules in the mesentery range. He then underwent an exploratory laparoscopy.

We used Open technique in periumbilical region for the first trocar (10 mm) and a 14-mmHg pneumoperitoneum was induced. Under direct vision, three other laparoscopic accesses of 10 mm, two in the right pararectal position, respectively above and below the umbilical, and one in the left sub-umbilical pararectal. A panoramic inspection of the abdominal cavity was carried out revealing the presence of a round mass arising in the colon trasversus mesenterium, which was first identified and then removed by means of an extraction bag. The postoperative clinical course was characterized by evacuation of faecis and gas for the two days after surgery. Three days after surgery we have noticed abdominal widespread pain and distention, with no evidence of peritonitis. We decided to replace the nasogastric tube and to perform an abdominal x-ray.

The abdominal x-ray showed a significant gaseous distention of the right colon till sigma, with air-fluid levels in

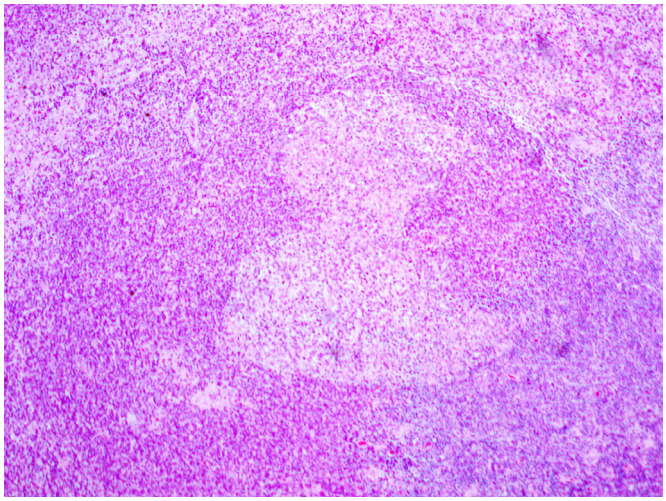

Figure 1. Castelmann's nodule: angiofollicular lymph node hyperplasia with prominent hyaline vascular lesions. Mantle zone present "onion skin" apparence. (Hematoxylin and eosin 4x).

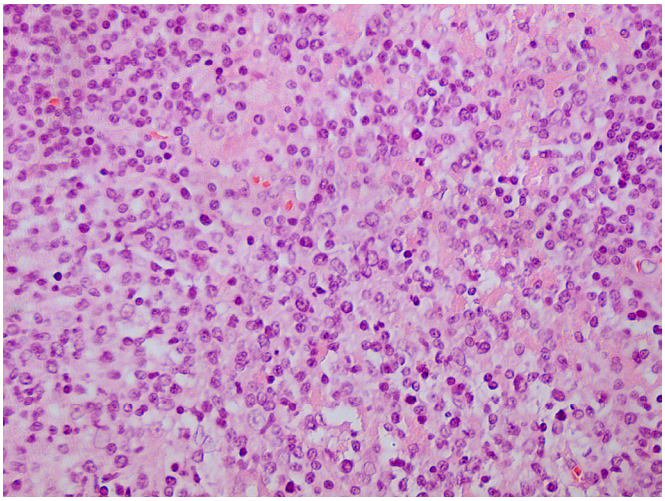

Figure 2. Castelmann's nodule: no atypical immunoblasts or plasmablasts are found. Prominent interfollicular vascular proliferation is observed. (Hematoxylin and eosin 20x).

the small intestine. Six days after surgery, we removed the nasogastric tube and the alvus was regular. The Patient started a liquid diet, returning to a normal diet eight days after surgery. In the 9th postoperative day the patient was discharged.

The histological diagnosis was Castelman's lymphadenopathy, hyaline vascular variant (Figs. 1, 2).

During Reumatological follow-up (last contact on September 2017), the regression of the lymphadenopathy was recorded, a low grade bi-weekly fever persisted (3–4 days lasting), until the introduction of Azatioprin 200 mg/die. The patient was found in good conditions.

Discussion

Castleman's disease is a very rare condition that may resemble more common disease. The prevalence of CD has not been estimated, but it has been calculated that the number of cases in the United States ranges from 30,000 to 100,000. Its incidence rate has not been reported in literature, although CD appears to be more common in the Asian population.[4] In a series of 16 cases in 1999 from the Memorial Sloan-Kettering Cancer Center (MSKCC), patients with unicentric CD were frequently accidentally discovered with symptoms mostly arising from mass compression.[8]

In 2005, Brusciano et al.[9] reported 330 cases of localized CD and about two thirds of the observed cases were localized in the mediastinum or hilus of the lung. The diagnosis is usually achieved only after surgical resection of the mass.

Currently, we have found more than 400 referred cases referred, with the localized form as the most frequent. The multicentric is the rarest variant, often related to immunosuppression and HIV infection.

CD can occur at all ages, where the majority of cases is asymptomatic (51%), especially in the localized form, which is often accidentally diagnosed.

As our case confirms, Unicentric disease is more common in the 3rd and 4th decade, whereas the multicentric form is more common in the 5th and 6th decade with no sex predilection.[1] Occasionally, when the lesion is large enough, compressive or constitutional symptoms may be present. It tends to occur in the third and fourth decade of life with a slight female predominance, with a median age of 35 years.[4]

Symptoms are related to the size of the affected lymph nodes (6 cm in average, ranging from 1 to 12 cm). In this case, chest or abdominal pain may occur due to the mass compression. In 30% of cases CD results in general systemic symptoms such as asthenia, fever, and weight loss.[6]

The multicentric variant is almost always symptomatic. Symptoms are fever, generalized lymphadenopathy, hepatomegaly and/or splenomegaly, POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy and Skin changes), Kaposi's sarcoma.[6]

The etiology of CD is unknown, but several studies highlight the role of HHV8 in the genesis of this disorder. HIV seropositive individuals appear to be at an increased risk for multicentric Castleman's Disease at a younger age due to the increased incidence of HHV- 8 infection.[1]

HHV8 is present in 60–100% of patients with CD and HIV, but it is also present in 20–40% in HIV negative patients.[10] The Epstein-Barr Virus, Toxoplasma and Mycobacterium tuberculosis have been considered to be involved in the genesis of some cases of variant PC CD. This disease could be a response to a chronic inflammatory process, an hamartomatous process, a state of immunodeficiency or an immune disorder.[3]

The diagnosis is based on the histological analysis of the lymph node using immunohistochemical methods.

The treatment of CD depends upon the type of the disease. Unicentric form is generally curable using surgical resection (with or without radiotherapy).[1] Radiation therapy has been used with mixed success in patients who are poor surgical candidates or Castleman's Disease, those with unresectable lesions.[2] At present, there is no consensus on the optimal management strategy for MCD. Successful treatment of MCD has been achieved using chemotherapy, with or without prednisone, given at the time of initial diagnosis.[1]

Surgery, radiotherapy, steroids, immunotherapy (interferon-α or anti-IL-6 antibodies), in combination with chemotherapy have all been used to manage the disease.[11] Complete surgical excision remains the most used treatment strategy for unicentric CD, which confers a cure rate approaching 100%.[12] Radiologic evaluation mainly involves computed tomography (CT), magnetic resonance (MR) imaging and positron emission tomography (PET/CT). The typical feature on CT scan is a well circumscribed

mass with soft tissue attenuation and rarely calcification. On MR imaging, these highly vascular tumors appear solid and have intermediate to high signal compared to muscle on T1 weighted images. Hyper intense signal is seen on T2 weighted images. PET scan is a useful modality for staging and monitoring response to chemotherapy.[13]

The treatment of CD depends upon the variety of disease. Unicentric form is generally curable using surgical resection (with or without radiotherapy).

CD must be included in the differential diagnosis of inhomogeneus nodules in the mesentery range, especially in rheumatological patients treated with immunosuppressive therapy, even though the definitive diagnosis can only be achieved by histological and immunohistochemical examination. Therefore, in the appropriate clinical setting, the diagnosis of CD must be considered, after investigating and excluding more common causes of lymphadenopathy or associated neoplastic processes. In cases of masses of an uncertain nature, laparoscopy must be considered the last diagnostic tool and the first treatment.

In the case of single location, the treatment of choice is surgical removal. In our experience, the laparoscopic approach is the most appropriate choice, if not contraindicated, because it allows to explore the entire abdominal cavity, early mobilization with a reduction of postoperative pain, and a shorter duration of hospitalization.

Disclosures

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

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