

Laparoscopic Management of a Unicentric Abdominal Mesenteric Localization of the Castleman Disease

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Abstract

Introduction: Castleman disease (CD) is a rare condition often discovered during routine checkups because of the lack of pathognomonic characteristics.

Case Presentation: A 55-year old male patient presented at the Emergency Unit for abdominal pain. The clinical examination revealed a soft abdomen, with slight pain in the left hypochondriac region, in addition to left-side lumbar pain. An abdominal CT scan identified a mesenteric mass in the left iliac fossa region. We performed a laparoscopic resection of the mass. The anatomopathological examination revealed a Castleman disease.

Discussion: Surgery can be curative in the unicentric form and IL-6 may play a prominent role in the development of the disease, causing prognostic and evolution factors.

Keywords: Laparoscopic Management; Castleman Disease; Giant Lymph Node Hyperplasia (GLNH)

Introduction

Giant lymph node hyperplasia (GLNH), also known as angiofollicular lymph node hyperplasia (ALN) or Castleman disease (CD), describes a rare condition first observed in 1954 and later refined in 1956 as part of Dr. Castleman's seminal work on the topic. The latter reported a series of cases exhibiting mediastinal masses, hinting at a lymphoproliferative disorder symptomatic of a thymic cancer. Yet, the absence of radiological elements characteristic and pathognomonic of the disease made it difficult to confirm such diagnostic [1]. A cytological examination is a very important for an effective diagnosis [2]. As the intentional diagnosis of Castleman disease is difficult, it is often discovered in accidental instances, during routine checkups or as a result of patients reporting specific symptomatology [3]. Sixty percent of the time, a mass is discovered in the thorax, against 14% and 11% in the neck and abdomen, respectively [2]. Additionally, at least 30% [4] of diagnosed patients went from perfectly asymptomatic to exhibiting symptoms ranging from acute coughing, pain, fever, anemia and in some instances, the discovery of a palpable mass [3,5].

Case Presentation

A 55-year old male patient presented himself at the Emergency Unit reporting left base-thoracic pain associated with dyspnea and productive 'wet' coughing. Pain radiated from left iliac fossa and lombal region, with no complaint of urinary pain. The patient suffered from asthenia and reported continued tobacco consumption, with a per-year estimate of 40 packs of cigarettes, and antecedent tuberculosis. Clinical examination revealed no abdominal sensibility, with slight pain in the epigastric and left hypochondriac region, in addition to left-side lumbar pain and left basal pulmonary crepitus. An abdominal CT scan revealed a mesenteric thickening in the left iliac fossa region

containing microcalcifications measuring 41 x 32 mm in axial and a loss cleavage between the mass and intestinal wall, hinting at a Gastro Intestinal Stromal Tumor (GIST) (Figure 1). The blood test reveals no anomalies. Exploratory laparoscopy performed in elective condition, along with mesenteric mass resection. The anatomopathological result was positive for Castleman disease.

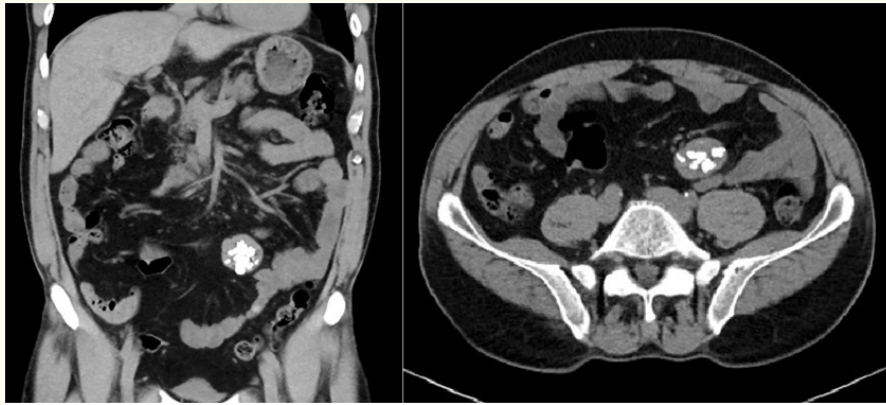


Figure 1: Abdominal CT scan who reveals a mesenteric calcified mass.

Discussion

The radiological, cytological and clinical means are used to further differentiate between the disease's two clinical identities [5,6]: (i) Unicentric Castleman Disease (UCD), solitary and localized, involving a single site for which the removal of the lymph node is curative [1]. Multicentric or systemic form of Castleman's disease (MCD), which is associated with the involvement of multiple lymph nodes. In 100% of cases, systemic symptoms are observed, including fever or anemia, and treatment determined using symptomatic scales [5,6]. Other studies also identified an extra nodal form of Castleman disease, arising from tissue other than lymph nodes or from sites which normally contain no lymphoid tissue such as a forearm muscle or cutaneous mass [7,8]. From an anatomo-pathologic standpoint, the existing research domain distinguishes three types of tissue lesions, depending on the type of follicular alteration of the nodes, or quantification of interfollicular plasma cells [7,9]. The most common tissue lesion called 'hyaline vascular' variant, primarily associated to the unicentric form of the disease, is characterized by hyperplastic germinal centers in the follicular region, in an onion-like arrangement or 'lollipop' appearance. The other type 'plasma-cell' variant, whose symptoms are harder to systematically diagnose, can be identified by an increase in plasm oblasts or plasmatocyte-like populations in the germinal center and/or paracortical region. Lastly, a mixed variant exists, pathologically classified as both hyaline vascular and plasma cell type [1,7,9].

An anatomopathological examination of the smooth surface module, weighing 30g and measuring 50 x 40 x 35 mm, further hinted at Castleman disease. Notably, the presence of a thickened mantle, in an onion-ring or lollipop arrangement, centered around transfixiant vessels, as well as two to three germinal center within a same follicle, known as 'twinning'. The diagnosis for UCD, the 'vascular hyaline' type, is made. Testing for B-cell monoclonality by PCR found no monoclonal rearrangement of IgH and IgKappa, excluding a lymphoproliferative disorder.

Operative technique

Insertion of a 12 mm trocar in peri-umbilical position and two 5mm trocars in the left iliac fossa region and left flank following the creation of a pneumoperitoneum. Exploratory laparoscopy revealed an oval mass, circumscribed and turgescient, found in the mesentery. Aciform incisions around the tumor, associated with bed-specific hemostasis, enabled complete extraction of the mass (Figure 2). Surgical access to the mass must be done carefully, due to significant vascular proliferation and endothelial hyperplasia, which may cause bleeding [2].

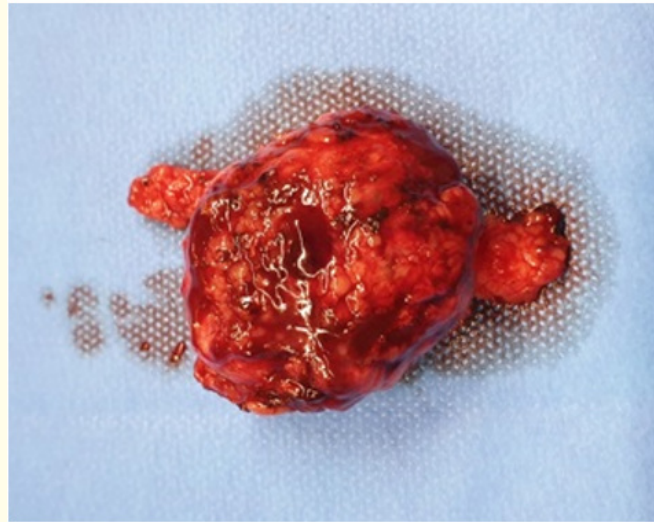


Figure 1: Abdominal CT scan who reveals a mesenteric calcified mass.

Etiologic diagnosis

Castleman disease is still a more or less unknown etiology, affecting males and females in equal numbers [10]. Studies have highlighted a strong correlation between HIV-positive patients surr infected by HHV-8 and an increase in diagnosis of MCD with plasmablastic cell type. It is associated with an increase in IL-6 expression by cells in the germinal center, as well as higher seric levels in patients with plasma cell variant [11-13]. The high levels of B cells associated with MCD are possibly due to an increase in levels of interleukin 6, one of the factors known as responsible for the differentiation of B-cells into plasma cells [2,12]. As such, infectious etiologies from HIV, HHV-8, Epstein-Barr virus, to toxoplasmosis or Mycobacterium tuberculosis cannot be excluded [3,4,14].

Advances and available treatments

The two identities of Castleman disease differ from a clinical standpoint, and hence also in their treatment. Surgical treatment is the Gold standard for treating UCD [15], with excellent results averaging a 100% remission rate according to studies from 1986 to 1997 including post-op follow-up at 12 and 76 months [3,4,16]. Radiotherapy has also proved beneficial and led to a decrease in ganglion size in MCD and UCD patients, with doses varying from 1800 to 4300 centi-gray (cGy) [4]. In the case of an unresectable mass with acute systemic manifestations, radiotherapy treatment is gaining momentum although less popular than surgical treatment [4,17]. In the case of symptomatic MCD, radiation treatment was also effective, relative to treatment combining chemotherapies or surgical treatment with partial resection of damaged tissue. Complete remission is possible, assessed against criteria such as the absence of observed pathology following radiography [14]. Patients with the plasma cell variant demonstrate higher seric levels in IL6 and a higher expression level marked in germinal center cells of the node, with no increase in other cytokine levels [17,18]. Such increase has been shown to significantly guide final prognostic [19]. (19) IL-6 is involved in the maturation of B-cells into plasma cells that secrete immunoglobulins. Any interference in their levels contributes to most MCD symptoms, suggesting they may play a prominent role in the development of the disease, causing prognostic and evolution factors. Consequently, targeted therapies incriminating anti-IL 6 antibodies significantly improve quality of life [18,19].

Conclusion

Castleman disease is difficult to diagnose due to lack of knowledge and pathognomonic characteristics. In term of treatment, surgery provides the cure for the UCD, but the recidive can occur even after a total resection. Radiotherapy and targeted therapies based on humanized anti IL-6 receptor and anti-IL-6 antibody's, preferably used when surgery shows no efficacy but seems to have benefits on survival and life quality.

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