Unicentric Mesenteric Castleman’s Disease- A Diagnostic Quandary- A Case Report

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ABSTRACT
Castleman’s Disease (CD) is a rare lymphoproliferative disorder and a mesenteric location is unusual. The unicentric variety is more difficult to diagnose clinically and it requires a histopathological confirmation. The excision biopsy itself will help both in the diagnosis and in planning the treatment. Here, we are presenting a case of unicentric mesenteric Castleman’s disease which we suspected to be lymphoma.

Key Words: Castlemans disease, Unicentric, Mesenteric

INTRODUCTION
Castleman’s Disease (CD) is a benign, rare, lymphoproliferative disorder which was first described by Dr. Benjamin Castleman in 1956. It is also known as angiofollicular lymph node hyperplasia, angiofollicular lymphoid hyperplasia, giant lymph node hyperplasia, lymphoid hamartoma, benign lymphoma or follicular lymphoreticularoma. This disease usually occurs in the mediastinum, lung, neck, axilla, pelvis and the retroperitoneum. But a mesenteric CD is rare. The diagnosis is difficult but the management depends on whether it is of the multicentric or the unicentric (localized) type.

The mesenteric Castleman’s disease has to be differentiated from other benign or malignant mesenchymal lesions of the mesentery like stromal tumours, leiomyomas, leiomyosarcomas and fibromas. It is also associated with a number of malignancies, which include Kaposi’s sarcoma, non-Hodgkin’s lymphoma, Hodgkin’s lymphoma, and the POEMS syndrome. We are reporting a case of mesenteric CD in a middle aged man.

CASE REPORT
A middle aged man was admitted with pain in the abdomen which was there since 2 months. On physical examination, a tender, oval shaped, palpable mass was found in the left infraumbilical region, which measured about 4cm. On admission, the investigations showed microcytic hypochromic anaemia (Hb%-6.2 G% and PCV-19.3%), proteinuria with elevated renal parameters (urea- 84 mg/dl and creatinine -5.1mg/dl) and chest X-ray with no radiological abnormalities. An abdominal ultrasound which was done, revealed a hypo echoic mass lesion in the left infraumbilical region. On further investigation with CT of the abdomen and the pelvis, a well-defined, soft tissue mass was seen in the mesentery, with internal foci of calcification and multiple enlarged mesenteric lymph nodes. The radiologist gave an impression of lymphoma [Table/Fig-1] and suggested fine needle aspiration cytology (FNAC)/biopsy for its confirmation.

CT guided FNAC showed the features of Non-Hodgkin’s lymphoma. However, the pathologist suggested a histopathological study of the mass for a confirmative opinion.

A minilaparotomy was done, which showed that there was a hard dark brown coloured mass in the mesentery which measured surgery section [Table/Fig-2]: Specimen Photo

[Table/Fig-1]: Ct Scan Showing Well Defined Soft Tissue Mass in the Mesentery with Internal Foci of Calcification

[Table/Fig-2]: Specimen Photo
The hyaline vascular type exhibits a prominent proliferation of the small hyalinized follicles, with a marked interfollicular vascular proliferation [Table/Fig-4]. The plasma cell type (10-20%) has hyper plastic germinal centres and sheets of plasma cells in the interfollicular region, with the proliferation of the blood vessels and the persistent sinuses, while a small per cent has a mixed histologic appearance with the features of both the hyaline vascular and the plasma cell subtypes.

Radiologically, the findings of CD are not specific and the radiological studies without histopathological reports will not give a definite diagnosis. CT scans can show a well defined soft tissue density, and the hyaline vascular type is more contrast enhanced than the plasma cell type.

Unicentric CD is characterized by the growth of lymph nodes at a single site with few (associated with the pressure symptoms) or no symptoms. Excision of the lymph nodes is the gold standard treatment, although there are no reported cases of recurrence [4]. There was a case of unicentric mesenteric CD which was of the hyaline vascular type, which presented with chronic non-iron deficient anaemia, which resolved after an effective treatment of the CD [5].

Unicentric Castleman’s Disease (UCD) is most often an isolated benign lymphoproliferative disorder of young adults, that is unassociated with the HHV-8 infection and that is generally curable with surgical resection. The largest published series included 81 cases; approximately 90 percent were of the hyaline vascular variant 3. UCD may be associated with an increased risk of lymphoma. Multicentric CD is always symptomatic as a result of the elevated Interleukin-6 production and is associated with asthenia (65%) weight loss (67%) and fever (65%) [6] and with the POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal proteinuria and skin changes).

The first case of multicentric CD was reported in 1978 [7]. Multicentric CD is usually a plasma cell variant and it is a systemic disease with generalized peripheral lymphadenopathy, hepatosplenomegaly, frequent fever, and night sweats. The multicentric type is associated with AIDS and the human Herpes virus 8-related infections. Many patients have been reported to die of the progressive disease with multi-organ system failure. No standard treatment for multicentric CD has been advocated. But a combined treatment of surgery, chemotherapy and corticotherapy is being tried [8,9].

The symptoms of Hodgkin’s disease and lymphoma can mimic those of Castleman’s disease. In Hodgkin’s disease, the symptoms may include fever, night sweats, weight loss, and/or enlarged or swollen lymph nodes. The tumours occur most often in the chest, stomach, or the spleen. Hodgkin’s disease is usually progressive and it may spread to involve the lymph nodes which are located in other areas of the body. Malignant lymphomas generally occur in the chest and/or the stomach. The symptoms which are common to all forms of malignant lymphomas include fever, excessive sweating at night, weight loss, and/or an enlarged liver and/or spleen (hepatosplenomegaly). Both the conditions can be evaluated and differentiated by a histopathological confirmation.

REFERENCES


The aetiology of CD is not properly known and is characterized by an lymphoid hyperplasia which mimicks lymphoma. It involves the hyper proliferation of certain B cells which produce cytokines. Castleman’s disease can be classified as a) unicentric or multicentric, based on the clinical and the radiological findings, b) as of the hyaline vascular or plasmacytic or mixed cellularity variety, based on the histopathology and c) as HIV negative or HIV positive based on the HIV status of the patient. Pathologically, CD can be of a hyaline

CD was first described by Dr. Benjamin Castleman in the year 1956, when he studied a group of patients with large thymoma like masses in the anterior mediastinum [1]. Although the involvement of CD is commonly seen in the mediastinum, neck, axilla, the shoulder region, the pelvis, pancreas, nasopharynx and the retroperitoneum, mesenteric CD is rare. This disease is more common in adults but it can be seen in children also [2].

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