Case Report

Castleman disease as an unusual cause of ascites: Case report

Ahmed Ramadan 1, Seham Seif 1, Ashref Elfakhry 1, Ahmed Eladl 2, Nora Shaband 3, Abdelfattah Abdalla 4
1 Internal Medicine Department, Faculty of Medicine, Mansoura University, Egypt. 2 Pathology department, Mansoura university, Mansoura, Egypt. 3 Nuclear medicine and oncology department, mansoura university, Mansoura, Egypt. 4 General surgery department, alazhar university, Egypt.

Abstract
Castleman disease (CD) is a collection of rare lymphoproliferative illnesses with similar lymph node histological findings. Castleman disease most commonly manifests with a hyperenhancing lymph nodal mass and should be considered in the differential diagnosis of lymphoma, metastatic adenopathy, infections or autoimmune diseases result in adenopathy. This disease includes a range of pathologic variants, including the Unicentric hyaline vascul type, plasma cell type and multicentric type and Castleman disease linked with human herpesvirus 8.

Introduction
Benjamin Castleman first described Castleman disease (CD), also known as lymphoid hamartoma or angiofollicular lymph node hyperplasia, in 1954. It refers to a group of uncommon lymphoproliferative illnesses with similar lymph node histopathological characteristics. lymphomadenopathy, splenomegaly, hepatomegaly, anaemia, skin and lung lesions, fluid accumulation, haemorrhage, infection 1. Historically, CD has been classified in two general forms, unicentric (localized) or multicentric type (systemic). The unicentric CD (UCD) form refers to 75% of all cases, with histological features of either hyaline vascular proliferation (90%) or mature plasma cell proliferation (10%). Unicentric CD usually comprises a single lymph node region, showing characteristic “Castleman-like” histopathologic changes and mostly found in the mediastium and abdomen. Multicentric CD (MCD) is sub classified to human herpes virus 8 (HHV8 or Kaposi Sarcoma virus 8 (mainly diagnosed in inpatients with HIV infection) and idiopathic MCD (iMCD) with histological features of both hyaline vascular and plasma cell proliferation with relatively preserved nodal architecture 1-3. Inflammatory manifestations are usually slight in UCD and frequently dissolve following surgical removal of the lymph node4; however, both iMCD and HHV-8-associated MCD are characterized by multifocal lymphadenopathy with a variety histopathology and episodic systemic inflammatory symptoms 4-8. The presenting features included, fever, sweating, fatigue, cachexia, generalized lymphadenopathy, splenomegaly, anasarka, cytopenias, and hypoalbuminemia 9-21. Recently, the Castleman Disease Collaborative Network (CDCN) proposed a classification system retaining the UCD vs MCD nomenclature, but further dividing MCD by etiological driver (HHV8-associated MCD [HHV8-MCD]; POEMS-associated MCD [POEMS-MCD]; iMCD) and within iMCD by phenotype, iMCD-TAFRO, and iMCD-not otherwise specified (iMCD-NOS) 12. Here, we report a case of ascites related to CD in peritoneal cavity.

Case Presentations
Male patient aged 24 years old, single with no special habits of medical importance complaining of sudden onset of ascites discovered accidently. He had no history of medical disease or surgical operations. He denied any recent travel and no sick contacts. The physical examination was totally normal except mild to moderate ascites. Laboratory investigation showed that, hemoglobin was 14.3 g/dl, platelets were 170x10^9/cm3, WBCs were 7.07 10^9/cm3, serum creatinine was 0.7mg/dl, serum albumin was 3.7 g/dl, total bilirubin was 0.6 mg/dl, AST was 13U/L, ALT was 12U/L, INR was 1.3 , CRP was negative, serum ferritin was 115 and serum thyroid stimulating hormone was normal. The patient had negative virological markers (Anti HCV antibody, HBsag and HIV1 and 2). Also, autoimmunity makers comprising, antimitochondrial antibodies (AMA) and anti-liver-kidney microsomal (ALKM) antibody were negative. ESR 1st hour was 5 mm/h and 2nd hour was 6 mm/h. Tumor markers (CA125, CA199, CEA, AFP, PSA) were unremarkable. Ascetic fluid analysis (SAAG was 0.9 , 96 % lymphocytosis , ascetic fluid ADA was negative, PCR for TB was negative, Negative ZN stain, glucose was 81 mg/dl, LDH was 148 U/l, protein was 6.2 g/dl. Ascetic fluid cytology showed mild lymphocytic and negative for malignant cell.

Post contrast pan CT (brain–neck-chest-abdomen and pelvis) showed mass in the right iliac fossa and bilateral enlarged axillary LN largest 12*7 mm and moderate to marked pelvi-abdominal ascites. Echocardigraphy was normal and Bone marrow deposits. Bone marrow aspiration showed no infiltration.

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* Corresponding author. email: Dr. Ahmed Ramadan Email: ahmedramadanabas86@gmail.com

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Laparoscopic reassessment demonstrated mass in the right iliac fossa that was biopsied for histopathological examination. The pathological examination showed atypical diffuse lymphoplasmacytic proliferation with occasional hyperplastic lymphoid follicle of unclear nature possibly castelman disease, associated with omental and peritoneal involvement fluid with mild reactive changes. Immunohistochemistry showed that, CD 20 showed moderate number of positive B-lymphocyte, CD 3 showed moderate number of positive T-lymphocyte, CD 138 showed FEW scattered positive plasma cells, Calretinin showed positive irritated surface mesothelial cells. Lastly, the patient was diagnosed with CD.

A, B, C: Florid mesothelial cells hyperplasia with papillary formations. The proliferation is mainly involving the mesothelial surface with focal limited involvement of the underlying tissue which shows dense mixed chronic inflammatory cells infiltrate. No detected invasion.D: Calretinin immunostain with positive cytoplasmic reaction in mesothelial cells. E: WT-1 immunostain with positive nuclear reaction in mesothelial cells.F: Cytokeratin immunostain with positive cytoplasmic reaction in mesothelial cells.

Discussion

In this case we found non-cirrhotic ascites in patient not complaining from other causes of ascites such as renal or cardiac causes. So when routine tests fail to disclose the source we progress to laparoscopy and according to histological examination we diagnosed this rare case of ascites to be Castleman disease related ascites. Castleman disease (CD) is a heterogeneous set of illnesses with a wide lymph node histological spectrum as well as systemic symptoms like fever, weight loss, and exhaustion, which are triggered by interleukin-6 (IL-6) and other cytokines. UCD, which involves a single enlarged lymph node or multiple enlarged lymph nodes within a single lymph node station, was first described by Castleman and Towne in a patient with a mediastinal mass in 1954 and soon thereafter by Castleman et al1, 2. Castleman disease usually progresses slowly, with lymph node enlargement occurring gradually. Patients can be diagnosed at any age (range 2-84 years), with a median age of diagnosis of 34 years and probably a slight majority of female patients. There are no known epidemiological factors that predispose to the development of UCD. The incidence of UCD has been estimated at 16 to 19 per million in the US population, which translates to 5000 to 6000 patients per year. UCD is virtually always HHV-8−, but rare positive cases have been reported, and these should be categorized and managed as localized HHV-8−associated CD. UCD is frequently diagnosed by chance, and lymphadenopathy is frequently asymptomatic. Some people, however, develop symptoms as a result of compression of important structures (such as the airways, neurovascular bundles, or ureters), while others develop iMCD-like inflammatory syndromes. The mediastinum (29%) was the most common location of involvement in a large series of UCD patients, followed by the neck (23%), belly (21%), and retroperitoneum (21%). (17 percent) UCD can also occur in the axilla and inguinal regions as well as orbits, nasopharynx, and small bowel. The mechanisms that was suggested for ascites development in this case may be compression of swollen lymph nodes in the abdominal cavity and retroperitoneum leads to obstruction of the lymphatic vessels and fluid leakage especially we donot found hypoalbuminemia or abnormal renal function in this. The first choice of treatment for UCD is surgical resection whenever possible. Complete surgical resection can cure almost all symptoms and return to normal laboratory abnormalities. The consensus guidance of van Rhee et al outlines the recommendations for the treatment of MCD, including Siltuximab, Tocilizumab, Rituximab, corticosteroids, immunosuppressive agents, etc., but the efficacy is worse than UCD. In 2017, a publication, based on the consensus work of multiple pathologists and clinicians to establish consensus diagnostic criteria for iMCD, was made.

References


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