ISSN: 2573-9565

Case Report

Journal of Clinical Review & Case Reports

Unicentric Castleman's Disease in a Man Originally Diagnosed with Sarcoidosis: A Case and Analysis of the Key Differences in Diagnosis

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Submitted: 03 Apr 2019; **Accepted**: 08 Apr 2019; **Published**: 18 Apr 2019

Introduction

Castleman Disease (CD) is a lymphoproliferative disorder characterized by enlarged hyperplastic lymph nodes with regressed follicles surrounded by expanded mantle zones of small lymphocytes, and interfollicular vascular proliferation in the hyaline-vascular type. There are two types: unicentric and multicentric. Unicentric, hyaline-vascular type of Castleman's disease can be treated successfully with complete surgical resection with monitoring for reoccurrence. Here we report a case of a patient originally diagnosed with sarcoidosis who was found to have Unicentric Castleman's disease.

Case

A 30 year-old incarcerated African American male with a history of shortness of breath for the past five years presents with acutely worsening dyspnea for two days prior to arrival to our hospital. The patient was admitted due to worsening dyspnea, chest pain, and non productive cough. The patient had been diagnosed with "sarcoidosis" on biopsy in 2013 and has been on steroids since then with some improvement until recently with no relief with his rescue inhaler. On admission the patient had chest computed tomography performed showing hilar and mediastinal adenopathy with diffuse airspace and ground glass opacities through the right lung. Imaging was most significant for total pulmonary artery compression which was secondary to a 3.9x 3.3 cm nodal mass in the right hilar region. There was also a confluent conglomeration of sub-carinal nodes measuring on the order of 3.2x2.8 cm. Laboratory testing was significant for increased IL-6,CH50, C3 and C4 complement. A ventilation perfusion scan showed no perfusion to the right lung. Pulmonary function testing (PFT) revealed decreased FEV1/FVC ratio and decreased DLCO when compared to previous testing. Cardiothoracic surgery was consulted and patient underwent mediastinoscopy with a biopsy showing histopathology and immunohistochemistry findings of giant lymph node hyperplasia and angiofollicular lymph node hyperplasia. His large coalescing mediastinal lymphadenopathy, coupled with imaging findings, inflammatory markers, and biopsy report confirmed the diagnosis of Unicentric Castleman's disease as opposed to the previous diagnosis of Sarcoidosis. After lymph node biopsy and removal patient was lost to follow and refused any further work up.

Discussion

Epidemiology and Pathophysiology

Castleman disease (CD) was first discovered in 1954 and is characterized in two specific subtypes: unicentric CD (UCD) and multicentric CD (MCD). The overall incidence of CD is estimated at 25 cases per million with 23% representing MCD and the rest comprising UCD.

The disease is distributed evenly between males and females although it is seen more in males in the HIV population. UCD usually affects the mediastinum and presents as a solitary lymph node mass, where MCD affects multiple lymph node sites predominantly the cervical region.1There has since been a link seen with HIV positive MCD and HHV-8 with increased IL-6. The association is thought to lead to angiogenesis by up regulation in vascular endothelial growth factor (VEGF). Anemia is also seen in MCD and is thought to be caused by anemia of inflammation secondary to IL-6 leading to an abundance of hepcidin.

Diagnosis

Due to the scanty amount of cases, awareness about the disease is pivotal to secure a diagnosis. There will be unifocal or generalized lypmadenopathy, splenomegaly, and constitutional symptoms. Laboratory findings include anemia, thrombocytopenia, hypoalbuminemia, polyclonal hypergammaglobulinemia and acute phase reactants. Elevated IL-6 and VEGF is sometimes also seen.2However, diagnosis is based on histopathological findings which reveal three specific subtypes:

1. Hyaline vascular type which has prominent vascular proliferation and hyalinization of the vessel walls.
2. Plasma cell type seen in UCD which shows aggregates of lymph notes with sheets of mature plasma cells in interfollicular areas
3. Mixed hyaline vascular plasma cell type seen in MCD showing a mix of hyaline vascular type and plasma cell type [1].

Treatment

For patients diagnosed with UCD complete surgical resection is preferred with survival rates of 95%. If there are compressive features debulking surgery is preferred. In MCD when surgery is not feasible cytotoxic chemotherapy as well as biologic medications directed against CD 20 and IL-6, bortezomib, and even antiviral agents have been used [2].

Sarcoidosis and Castleman Disease in the Literature

Due to the diversity of its presentation Castleman disease is a great

mimicker of sarcoidosis as well as other malignant disorders. In the literature we were able to find another case where sarcoidosis mimicked Castleman disease. In both their case and ours the lung lesion, accompanied by persistent enlarged hilar and mediastinal lymphadenopathy which, most importantly, showed a blunted response to glucocorticoids was the clue to diagnosis prior to biopsy. However, biopsy is essential in differentiating the two disorders. In their case of MCD the disease took 10 years to fully manifest it. Our case of UCD took 6 years to properly manifest itself which is a slower progression than usually seen in sarcoidosis which can also be a pivotal clue in diagnosisb [3]. There is also the possibility of both diseases occurring simultaneously. In a review of the literature there were three reports of CD being discovered on an incidental follow up chest x-ray after a diagnosis of sarcoidosis. The relationship between the two diseases is not completely understood. Malignant lymphoma is a known complication of pulmonary sarcoidosis as well as a complication of MCD but the two diseases have not been linked so case reports linking the two are imperative [4]. Regardless, a pathological diagnosis is pivotal. In cases of MCD superficial lymph nodes can be used but if there are none thoracotomy or mediastinoscopy may be necessary for biopsy [5].

Conclusion

Castleman Disease (CD) is a lymphoproliferative disorder that is rarely seen. This is the second case documenting Sarcoidosis mimicking it. It is important to note it has also been seen occurring simultaneously with sarcoidosis leading us to believe that CD could be a complication of sarcoidosis in the same way malignant lymphoma has been linked to both diseases. Finally, although case reports help in characterizing the clinical features of both diseases, pathological biopsy is necessary in differentiating the two diseases. Castleman disease can be treated successfully with complete surgical resection with monitoring for reoccurrence. In cases where surgical resection is not possible chemotherapy and immunomodulators are now being used to treat CD.

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