Diagnosis of Castleman Disease

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KEYWORDS

- Castleman disease • Diagnosis • HHV8 • POEMS • TAFRO
- Paraneoplastic pemphigus

KEY POINTS

- Castleman disease (CD) comprises a heterogeneous group of disorders that share pathology similarities but present with diverse clinical manifestations.
- Specific clinical signs and complications, including paraneoplastic pemphigus, peripheral neuropathy, TAFRO (thrombocytopenia, anasarca, fever, reticulin fibrosis, organomegaly) and POEMS (polyradiculoneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder, skin changes) syndrome, or human herpesvirus 8 (HHV8) infection, are important features of CD's clinical spectrum that should be recognized and identified.
- Evaluation of CD should include, besides pathologic evaluation with immunostaining, laboratory investigations as well as systemic imaging with PET/computed tomography, both to stage the extent of disease (unicentric vs multicentric) as well as for markers for follow-up.
- HHV8-related CD requires evaluation for the presence of Kaposi sarcoma and HIV infection and is associated with increased risk of lymphoma.
- Lymphoma and autoimmune connective disorders can present with Castleman-like lymph nodes pathology and need to be excluded.

The original description of Castleman disease (CD), corresponding to the presence of angiofollicular lymph-node hyperplasia with capillary proliferation, hyperplasia of lymphoid follicles, and cellular infiltration of plasma cells, was first reported in 1956 in a series of patients with few or no symptoms but solitary mediastinal lymph node enlargement.[1] Sixty years later, CD remains a rare condition that comprises 3 distinct entities that share pathologic similarities regarding germinal centers, follicular

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dendritic and plasma cell prominence, and vascularity within lymph nodes but are featured by specific clinical, pathologic, and biological abnormalities. The lack of specificity of most of the pathology and clinical findings observed in CD requires adequate criteria to diagnose CD and rule out differential diagnoses. Multicentric Castleman disease (MCD) is classically distinguished from unicentric Castleman disease (UCD) by the presence of systemic symptoms (fever, asthenia, pleural effusion, ascites), presence of lymph nodes in more than one region, hepatosplenomegaly, and important signs of biological inflammation, as well as a poorer prognosis. MCD comprises 2 subgroups: human herpesvirus 8 (HHV8) -related MCD[2,3] and idiopathic multicentric Castleman disease (iMCD) which is not associated with any known etiologic factor.[4] Here, the authors review the clinical and pathologic situations that should lead to diagnosis of CD.

To establish the diagnosis of CD and specify its subtype, a complete clinical evaluation associated with lymph node biopsy and biological and morphologic examination is necessary. Here, we review the clinical and pathologic situations that should lead to diagnosis of CD.

PRESENTATION

The presentation of CD is varied. In patients with UCD, presentation is quite often asymptomatic with accidental detection of visible or palpable mass (enlarged lymph node) or abnormal laboratory tests on routine or unrelated examination. Rarely, UCD may present with systemic symptoms or because enlarged lymph node may impede nearby organs. On the other hand, MCD often presents with fever, night sweats, weakness, severe fatigue, and anorexia accompanied by weight loss. These classic systemic symptoms of MCD, are considered mainly driven by interleukin-6 (IL-6). Patients may have symptoms associated with complications of CD, including cutaneous, neurologic, or autoimmune manifestations. Because no symptom or laboratory investigation is diagnostic of CD, the ultimate and often the first investigation is lymph node biopsy with careful histologic examination by an experienced pathologist.

HISTOPATHOLOGICAL EXAMINATION OF CASTLEMAN DISEASE

Three distinct subtypes of CD can be distinguished based on lymph node pathology examination (see **Table 1**). Benjamin Castleman's initial report corresponds to the hyalin-vascular subtype, which is the most common feature of UCD.[1,5]

In the hyalin-vascular subtype, the lymph node architecture is featured by lymphoid follicles with atrophic or "regressed" germinal centers often hyalinized and mainly constituted by residual follicular dendritic cells, and prominent mantle zones containing small lymphocytes are seen. The follicular dendritic cells are organized in concentric form and provide an "onion-skin" appearance. Sclerotic blood vessels are often

Table 1
Histologic and clinical subtypes of Castleman disease

Histologic Lesion	UCD	HHV8-Related MCD	iMCD
Hyaline vascular	++++	+/–	+
Plasmacytic	+	+++	++
Mixed	+/–	++	++

UCD is more often associated with the hyaline-vascular subtype, whereas MCD is more often associated with plasma cell and mixed subtypes. The presence of plasmablast is exclusively observed in the context of HHV8-related MCD.

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