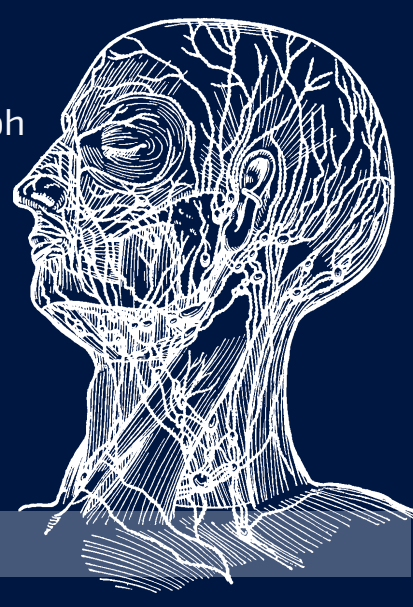
IDIOPATHIC MULTICENTRIC CASTLEMAN DISEASE

SYMPTOMS, DIAGNOSIS, & TREATMENT

Castleman disease is a group of rare disorders characterized by lymph node enlargement, specific microscopic changes to the lymph nodes, and a broad range of symptoms and laboratory findings. **Idiopathic multicentric Castleman disease (iMCD)** also known as HHV-8 negative MCD is diagnosed in patients with enlarged lymph nodes with Castleman disease features in multiple regions of the body and accompanying characteristic laboratory abnormalities, in the absence of HHV-8 infection.



Causes and Risk Factors

There are no known causes or risk factors for iMCD hence, the cause is said to be "idiopathic."

Symptoms

In iMCD, symptoms occur due to a hyperinflammatory response and tend to be more severe than unicentric Castleman disease symptoms. iMCD can progress to life-threatening multi-organ failure.

- flu-like symptoms: fever, night sweats, loss of appetite
- unintentional weight loss
- weakness, fatigue
- enlarged liver or spleen
- peripheral neuropathy (burning or numbness in hands or feet)
- kidney dysfunction
- edema (swelling) or ascites (fluid accumulation in the abdomen)
- cherry hemangiomas or skin rashes

Diagnosis

- Lymph node biopsy: Diagnosis of CD requires an excisional lymph node biopsy that shows features of Castleman disease.
- “Excisional” means that the lymph node must be surgically removed entirely so that it can be specially prepared and evaluated by a pathologist.
- Imaging: In MCD patients, imaging reveals several regions of enlarged lymph node.
- Laboratory tests are usually abnormal, with anemia, elevated inflammatory markers (CRP, ESR), and abnormal liver and kidney function tests.

Treatment

- Treatment is focused on controlling inflammation and preventing flares.
- IL-6 inhibitors are recommended first line:
  - Siltuximab (Sylvant) is the **only FDA-approved treatment** for iMCD in the USA
  - Tocilizumab (Actemra) which targets the IL-6 receptor is approved for use in iMCD in many other countries; recommended when siltuximab is not available
- Other commonly used medications include:
  - Rituximab (anti-CD20 antibody)
  - Sirolimus (mTOR-inhibitor)
  - Cytotoxic chemotherapy
  - Steroids can help dampen down the inflammatory response but don't directly treat iMCD

Talk to your physician to about the best treatment plan for you.

Subtypes

There are two currently recognized subtypes of iMCD:

iMCD

iMCD-TAFRO

iMCD-TAFRO describes an aggressive clinical subtype of iMCD involving
Thrombocytopenia
Anasarca
Fever
Reticulin fibrosis
Organomegaly

iMCD-NOS

iMCD-not-otherwise specified (NOS) describes iMCD patients who do not meet the criteria for iMCD-TAFRO. Compared to iMCD-TAFRO, iMCD-NOS cases tend to have a less aggressive clinical course.

CDCN
CASTLEMAN DISEASE COLLABORATIVE NETWORK
CASTLEBANK CDCN
ACCELERATE CDCN

What can you do right now?

- **Become a member of the CDCN network:** cdcn.org/patients-loved-ones/patient-login
- **Enroll in ACCELERATE to donate your medical records for research:** cdcn.org/accelerate
- **Donate samples to CastleBank (blood, lymph node tissues, etc.) for research:** cdcn.org/samples
- **Join CDCN Connect to connect with other patients:** connect.cdcn.org

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