

Idiopathic Multicentric Castleman Disease Tweetorial

This CME/CMLE Tweetorial from Dr. David Fajgenbaum highlights key concepts, diagnosis, and guidelines on multicentric Castleman disease. It was originally posted on ASCP's Twitter channel at: https://twitter.com/ASCP_Chicago/status/1621614779344326656

Review the recap below, then claim your free CME/CMLE credit at: <https://bit.ly/CastlemanTweetorial>

This activity is funded by an independent educational grant from Recordati Rare Diseases Inc.

Multicentric Castleman disease (MCD) is a rare inflammatory/lymphoproliferative disorder involving a cytokine storm and enlarged lymph nodes with characteristic microscopic features.

Learn more at: <http://bit.ly/3DD5ieD>

MCD should be divided into 3 subtypes: MCD caused by uncontrolled HHV-8 infection (HHV-8-MCD), MCD caused by cancerous plasma cells (POEMS-MCD), & MCD with unknown cause (idiopathic, iMCD).

Learn more at: <https://bit.ly/3Y3CaVQ>

We'll focus primarily on iMCD. Symptoms of iMCD include fever, fluid accumulation, lymphadenopathy, anemia, and low platelets. Symptoms can be non-specific and may mimic other illnesses.

Learn more at: <http://bit.ly/3HXg5mt>

A definitive diagnosis of iMCD requires a biopsy of an enlarged lymph node that shows characteristic histologic features.

Learn more at: <http://bit.ly/3Dzxf6L>

Polling Question

For pathologists, when you suspect a patient has a form of Castleman disease, what do you write on the pathology report or do?

1. Castleman disease
2. Castleman disease- like histopathology
3. Ask a colleague for help
4. Not sure

Polling Results

| | |
|---|-------|
| Castleman disease | 16.7% |
| Castleman-like histopatho <input checked="" type="checkbox"/> | 83.3% |
| Ask colleague to review | 0% |
| Other (please comment) | 0% |

Diagnosis requires multicentric LAD, histopathologic confirmation, presence of 2+ clinical “minor criteria” & ruling out all exclusion criteria.

Go here for diagnostic criteria and images: <http://bit.ly/3JCOwQm>

Once diagnosed, treat iMCD first-line with anti-IL-6 therapy (siltuximab). In pts with severe disease who are progressing on anti-IL-6 therapy, chemo may be needed.

Treatment guidelines available here: <http://bit.ly/3I5Skzs>

Siltuximab is a monoclonal antibody that targets interleukin-6, a cytokine that plays a key role in the pathogenesis of iMCD. Siltuximab is approved to treat iMCD in the US and 40+ countries worldwide.

In patients with milder iMCD who do not respond to siltuximab, other therapies (sirolimus, rituximab, ruxolitinib, thalidomide) have shown promise and can be used.

Info on sirolimus by Dr. David Fajgenbaum available at: <http://bit.ly/3X2JjEJ>.

Info on an open trial of sirolimus at: <http://bit.ly/3RxgmiR>

Rituximab is a monoclonal antibody that targets CD20 and is used in the treatment of HHV-8-associated MCD. Plasma cell directed therapies are used for POEMS-MCD.

iMCD is a rare and complex disorder, so a multidisciplinary approach to diagnosis and treatment is essential.

Go to <http://bit.ly/40sBdlb> to get involved and connect with others.

Specialists such as hematologists, oncologists, and pathologists should be involved in the care of patients with iMCD.

Close monitoring and follow-up are important in the management of iMCD, as relapses and complications can occur.

Further research is needed. If you have iMCD or have a patient with iMCD, please encourage them to learn about joining the CD registry and/or donating samples here: <http://bit.ly/3RtZcCK>



STRONGERTOGETHER

MCD is a rare disorder, but with early diagnosis and appropriate treatment, the prognosis for patients can be improved.

Learn more through ASCP's new microlearning series and podcast on MCD at:

<https://www.ascp.org/content/learning/castleman-disease>

Claim your free CME/CMLE credit for participating in this activity here:

<https://bit.ly/CastlemanTweeetorial>