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The Necessity of a Multidisciplinary Roundtable in the Diagnostic Workup of Castleman Disease

Announcer:
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Dr. Casper:
This is CME on ReachMD, and I'm Dr. Corey Casper. Here with me today is Dr. Sudipto Mukherjee.

So, Dr. Mukherjee, once a clinician includes Castleman's disease in their differential diagnosis, what do they do next? Can you give us an overview of what a diagnostic workup entails for multicentric Castleman's disease and who is involved?

Dr. Mukherjee:
Thank you, Dr. Casper. This is a very important question because, as we know, there are several conditions that can mimic iMCD [idiopathic multicentric Castleman disease]. Based on the international evidence-based diagnostic criteria, the diagnostic workup of iMCD is a multistep process and requires a 3-part criteria.

The first part is the presence of both of the 2 major criteria. Number one is enlarged lymph nodes in 2 or more lymph node regions, either assessed by a CT scan of the neck, chest, abdomen, and pelvis or a whole-body FDG PET scan. And the second major criteria is the presence of specific histopathological features that are consistent with the iMCD spectrum. Now, for this histopathological examination, it is critical that this is performed on an excisional biopsy of the involved lymph node.

The second part of the criteria is presence of at least 2 of the 11 minor criteria, of which at least 1 has to be a lab criteria. These minor criteria include a mix of both clinical as well as laboratory features.

And the third part is exclusion of all conditions or diseases that mimics iMCD. And these conditions or diseases can be broken together into 3 main groups. Infection-related disorders: Particularly for iMCD, it is important to rule out HHV-8 infection, either by a blood PCR [polymerase chain reaction] or LANA-1 staining by immunohistochemistry of the lymph node, and HIV infection. Second group of conditions that mimic iMCD that needs to be ruled out is autoimmune or inflammatory conditions. And the third is malignancies.

So as you can see, based on the extent of the workup and the multistage process, it is not surprising that by the time a patient gets diagnosed with iMCD, multiple specialties have been involved in one way or the other, leading up to the diagnosis. And that could include any one of the different specialties, such as EMT, rheumatology, infectious disease, hematology, oncology, general surgery, and sometimes even internists or family practice physicians.

Dr. Casper:
Thanks, Dr. Mukherjee. So I think you really have highlighted what makes the diagnosis and workup of a patient with Castleman's disease so challenging, but also some of the recent developments that have been so important.

So first and foremost, it's so wonderful now that we have established diagnostic criteria. For a disease as complicated and

heterogeneous as Castleman's disease, the fact that there are diagnostic criteria that clinicians can refer to is wonderful. And you very eloquently pointed out that one must have 2 of the major criteria. That includes the enlarged lymph nodes and an excisional biopsy that shows changes in a lymph node consistent with Castleman's disease and negative for a virus like human herpes virus-8. So those are our major criteria.

You then really nicely explained how there have to be at least 2 minor criteria, one of which has to be a laboratory criteria. And that's, again, because some of the minor criteria could be subjective. And this constellation of major and minor criteria really guides clinicians towards making a diagnosis. But to finally establish that diagnosis, also, other common diseases must be excluded, as you mentioned, things like infectious, autoimmune, and malignant diseases.

You did a really nice job of talking about the wide constellation of clinicians that often are involved in a diagnosis. The combination of maybe a general practitioner or a family practitioner, an infectious disease, cancer, or rheumatology specialist, and of course, the importance of the general surgeon, and the pathologist who helped to obtain these tissues and diagnose them to make the definitive diagnosis.

So no doubt a complicated disease to diagnose and work up. And a wide variety of clinicians are often called to make that diagnosis. But taken together, I think we have a firm road map now for the appropriate workup to make the diagnosis of Castleman's disease.

Thanks again for chatting with us. This has been a great micro discussion. Unfortunately, our time is up for today, and thanks for listening.

Announcer:
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