Transcript Details
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Diagnosing Castleman Disease: Key Considerations

Announcer:
You're listening to *Project Oncology* on ReachMD. On this episode, Mateo Bustamante will discuss how we can optimize the diagnosis of idiopathic multicentric Castleman disease, or iMCD for short. Mr. Bustamante is a Senior Clinical Data Analyst for the Center for Cytokine Storm Treatment and Laboratory at the University of Pennsylvania's Perelman School of Medicine. He also presented a session on this topic at the 2024 American Society of Hematology Annual Meeting. Let's hear from him now.

Mr. Bustamante:
Idiopathic multicentric Castleman disease is an incredibly rare and life-threatening disorder that affects around 1,300 newly diagnosed patients a year. And it impacts the body with these severe systemic inflammatory symptoms that can manifest in a variety of ways, from some moderate constitutional symptoms, such as fatigue, weight loss, unexplained fevers, and poor appetite, to more severe multisystem organ dysfunction that can result in death due to a severe cytokine storm that usually involves interleukin-6.

So my presentation at ASH focused on how the diagnostic criteria for idiopathic multicentric Castleman disease and using excisional biopsies speeds up diagnosis times for iMCD patients. We can break down iMCD patients into these two subtypes: the NOS, not otherwise specified subtype, and these are patients that usually have chronic moderate constitutional symptoms, and the TAFRO subtype. This is your severe iMCD subtype which stands for thrombocytopenia, anasarca, just fluid accumulation all across the body; the F is for fever or elevated C-reactive protein; R can be reticulin fibrosis in bone marrow or renal dysfunction; and O is organomegaly. And depending on the definition of TAFRO you use, that's either hepatomegaly, splenomegaly, or even enlarged lymph nodes. These two subtypes probably have very different diagnosis times due to the different clinical presentation. You have your severe multisystem organ failure compared to just moderate symptoms. And what we showed is historically, yes, these NOS, these moderate patients, they're taking significantly longer to get diagnosed than your TAFRO patients. Just looking at all time, not before or after diagnostic criteria, the TAFRO patients had a median diagnosis time of 35 days compared to nearly 120 days for NOS patients.

Now, when we're looking at the time period after the diagnostic criteria—and we limited our analyses to plus or minus five years to have a fair comparison time before and after—the TAFRO times are already short. They don't change much at all. But the diagnosis times for these NOS patients, these patients with moderate chronic symptoms, significantly reduces from 218 days before diagnostic criteria to 53 days after diagnostic criteria. Of course, there are so many other factors that have been going on during this time. Just Castleman disease research and groups like the Castleman Disease Collaborative Network have been spreading the word about Castleman disease, but I still think it's a really significant finding and just points to how useful these criteria are for physicians in diagnosing this incredibly rare disease.

So the potential consequences of delayed diagnosis on idiopathic multicentric Castleman disease patients—first of all, there's only one FDA-approved drug for iMCD. It is siltuximab. It is an interleukin-6 inhibitor. Unfortunately, it only works in a third to half of patients, but for the other half of patients, this could completely change their lives. And you can't know to administer siltuximab until you make the iMCD diagnosis, and that is why it's so important for the diagnosis to come quickly—so you can start the patient on the right treatment. So in situations where patients are started on the right treatment too late due to all their organs just having suffered too much damage, at that point, the medication can't save that person. So the diagnosis has to come quick so the doctors know to administer the right medication.

Also, the biggest challenges for iMCD specifically I would say is that so many diseases mimic iMCD, and iMCD just presents like so many other diseases, and that's why a major part of the diagnostic criteria is the exclusionary diseases that you must rule out that can mimic iMCD before you diagnose iMCD. And these disorders fall into three different categories: infectious-related disorders, such as

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EBV—you should be checking EBV titers—autoimmune diseases such as systemic lupus erythematous and RA, and then you have the neoplastic disorders that you have to exclude like your lymphomas and your multiple myelomas. When making the iMCD diagnosis, yes, you can have a lymph node that looks like iMCD and it might have the features of iMCD, but just be cognizant that you should still be checking for these other disorders that can appear like iMCD themselves.

Announcer:
That was Mateo Bustamante talking about the importance of early diagnosis for idiopathic multicentric Castleman disease, which he spoke about at the 2024 American Society of Hematology Annual Meeting. To access this and other episodes in our series, visit *Project Oncology* on ReachMD.com, where you can Be Part of the Knowledge. Thanks for listening!

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