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Climbing the Long, Winding Stairs to the Diagnosis of Castleman Disease

Announcer:
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Dr. Mukherjee:
Welcome to this CME on ReachMD, and I am Dr. Sudipto Mukherjee. Here with me today is Dr. Corey Casper and Jovanna, a patient who has been diagnosed with Castleman disease.

Let's start with hearing from Jovanna. Can you give us a brief overview of your clinical experience leading up to your definitive diagnosis with Castleman disease?

Jovanna:
Hi. Yeah. So as I mentioned before, I had a plethora of symptom that no one seemed to be able to pinpoint to one disease. I had abdominal swelling that made me look like I was in my third trimester of pregnancy. I walked into an ER, and they actually asked me if I was an active labor. During the 5 months that it took for me to get my diagnosis, I went to 4 different hospitals. And when I was at my most sick, they actually sent me home because they said that they had run every test that they could think of, and that they didn't have any answers for me. So at the time, they had done 10-plus tests, and they repeated some of those scans and tests multiple times. I felt totally hopeless and alone during this time period. There were no answers; I was getting more and more sick. I had a few doctors who apologized for not knowing what was happening. I had one doctor who quite literally threw up his hands and said, “I don't know what to do next.” And I had one doctor who told me that I felt so sick because I was always in bed, and that I needed to get up more and open the windows, and if I walked around, I would be fine. So I just felt this combination of confusion and anger and frustration, and I just felt like it would never end.

Dr. Mukherjee:
Thank you so much for sharing your clinical experience. And I'm not surprised. This is usually a very typical experience faced by several iMCD [idiopathic multicentric Castleman disease] patients.

Dr. Casper, why is it so challenging for clinicians to identify Castleman disease in patients like Jovanna?

Dr. Casper:
Yeah, thanks, Dr. Mukherjee, and thanks for sharing your story, Jovanna. I would say that this is a very common clinical presentation of Castleman’s disease. The symptoms are very nonspecific. So patients have fevers, night sweats, swelling, and organ dysfunction, which can happen in so many different diseases from infectious diseases to malignancies to rheumatic diseases. And because these presentations are so diverse and so nonspecific, it can make it very difficult for clinicians to narrow down on what actually is the cause of the patient's illness.

The unifying feature in Castleman’s disease, the one thing that brings all of these symptoms together is that the body makes too much of the cytokine interleukin-6. This is a signal that the body uses to do a number of different things like recruit immune cells or adapt to an

infectious threat. So these symptoms that we're seeing are all due to too much interleukin-6 production, but it can be hard to recognize.

Now, the other thing that makes a diagnosis of Castleman's disease difficult is that there are multiple different variants of this rare disease. It's bad enough that this is a rare disease that most clinicians only heard about a few times, maybe, in medical school and in their residency training. But the fact that there's multiple variants of it makes it even more challenging. At the very basic level, there's a unicentric, which is a single lymph node that's involved or lymph node chain, and multicentric, which means that multiple different lymph nodes are involved. In the multicentric Castleman's disease flavor of Castleman disease there are even subvariants within that. So there's a variant called idiopathic multicentric Castleman disease and that is one that has no known cause. There's also a variant of multicentric Castleman disease that is caused by a virus, human herpes virus-8. So it's important to understand the distinction between those 2 different variants of multicentric Castleman's disease. Then even within idiopathic multicentric Castleman's disease, there are subvariants. So there's a subvariant that's associated with POEMS, and another subvariant that's associated with TAFRO. Finally, there's the most common subvariant which is not otherwise specified. So that alphabet soup makes it very challenging for clinicians to keep this disease straight, and it also impedes diagnoses.

Finally, the fact that you can't keep straight that there is a single disease with a single treatment means that this is even more difficult to diagnose as well. So each of these different variants has a different way that you clinically approach it, and that can make a diagnosis challenging as well.

Dr. Mukherjee:
Thank you, Dr. Casper, for your concise yet comprehensive view of the clinical features and pathophysiology of Castleman's disease.

Well, this has been a great bite-sized discussion. Unfortunately, our time is up. Thanks for listening.

Announcer:
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