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Time needed to complete: 17m

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Hiding in Plain Sight: Could This Be Castleman Disease?

Announcer:
Welcome to CME on ReachMD. This episode is part of our MinuteCME curriculum.

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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 and Jovanna, a patient who has been diagnosed with Castleman's disease.

Let's dive right in with a discussion of the clinical burden of Castleman's disease and the impact that a delayed diagnosis has on our patients. So starting with Dr. Mukherjee, what are some of the challenges in identifying and diagnosing patients with Castleman's disease?

Dr. Mukherjee:
Castleman disease patients can have myriad presentations, depending upon the particular type of Castleman's disease and the extent of underlying inflammation. And that determines how much of a clinical burden each patient will experience through their clinical journey.

For example, for patients with idiopathic multicentric Castleman disease, or iMCD, the focus of this discussion, presentation can vary widely, ranging from a mild flu-like illness, to commonly seen constellation of findings such as fevers, night sweats, pain, lymphadenopathy at multiple sites, enlarged liver or spleen, generalized fluid accumulation in the body or in the belly, to severe sepsis-like picture with multi-organ failure and death. These symptoms can present abruptly or over a period of time with multiple hospitalizations and multiple specialty visits in the interim.

Diagnosis is very challenging as it is a multi-step process that requires excisional biopsy of the involved lymph nodes, demonstrating the specific histopathologic features, meeting certain lab criteria and clinical criteria, and importantly, exclusion of a variety of conditions that mimics iMCD.

Two of the most frequently encountered challenges in diagnosis is reliance on fine-needle aspiration biopsy of the involved lymph nodes for histopathological examination and some level of lack of awareness of the diagnostic criteria. But early diagnosis is critical as it leads to timely initiation of IL-6-directed therapy.

Now, I will turn over to Jovanna so that she can share with us her experience leading up to her diagnosis of Castleman's disease.

Jovanna:
Thanks, Doctor. So my first symptom was an incredible pain that I had in my abdomen, and that led to a myriad of other symptoms popping up as I got more and more sick. I had horrible night sweats. I was sweating through my clothes and my sheets every single night. My body began to swell and my abdomen got so swollen and full of fluid that I looked like I was 9 months pregnant. I was always nauseous or vomiting. And on my third hospital visit, they found out that my kidneys were functioning at 14%. By that time, I was so frustrated with going in and out of the hospital with no answers that I was fighting to go home. I ended up on dialysis and a PCA [patient-controlled analgesia] pump that gave me pain medication every 8 minutes because I wasn't able to function by myself because I was in

such incredible pain. I cried all the time, because the pain was getting worse and worse, and no one had answers for me. At first, most of my doctors thought that I had lymphoma because of all the lymph nodes that I had all over my body that were an increased size, but nothing else checked the boxes of lymphoma, so it became kind of an infuriating puzzle for everyone, including myself.

Dr. Casper:
Thank you for sharing your story with us, Jovanna. That sounds incredibly difficult and frustrating, and I can only imagine how frightening that must have been for you.

You know, Jovanna and Dr. Mukherjee both, I think, provided us with a very consistent and, I think, all too common portrait of what Castleman's disease looks like in a patient when it first presents. From Jovanna, we heard that there are a wide variety of symptoms and these include pain, night sweats, swelling, nausea and vomiting, and organ dysfunction. And again, the experience that Jovanna had with multiple attempts – 3 hospitalizations before a diagnosis was reached, is very common in patients with Castleman's disease.

From Dr. Mukherjee, again, we learned that it's this myriad of symptoms and the multistep aspect of how to make definitive diagnosis, the lack of familiarity that clinicians have with the guidelines around making a diagnosis all lead to delayed diagnoses.

So this has been a great initial bite-sized discussion. Unfortunately, our time is up for now. Thank you all for listening, and I look forward to exploring this further in subsequent sessions.

Announcer:
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