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Pinpointing CIDP: Diagnosis Challenges and Misdiagnosis/Delayed Diagnosis of CIDP

Announcer:
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Dr. Allen:
This is CME on ReachMD. I'm Dr. Jeffrey Allen. Joining me today is Dr. Nick Silvestri. Nick, thanks for joining us.

What are some of the diagnostic challenges that contribute to misdiagnosis of CIDP?

Dr. Silvestri:
Yeah. I mean, I think that's a great question, Jeff. I think that you and I can both agree that CIDP is probably one of the more overdiagnosed diseases that we see. I'm sure both of us get a lot of referrals for second opinion on CIDP and we figure out that the patient unfortunately doesn't have CIDP.

I think that one of the biggest pitfalls is we don't really have a diagnostic biomarker, right? Unlike a disease like myasthenia gravis where the majority of patients will have positive antibody testing, which really helps us make the diagnosis, that's just not the case in CIDP. And so we rely on our clinical history and our exam and then we use electrophysiology, as we've previously discussed, to really help solidify that diagnosis. But I think electrophysiology can be fraught by a number of different problems.

First and foremost, there are technical issues. If nerve conduction studies, in particular, aren't performed with technically accurate methods or procedures, conduction studies that would be otherwise normal or perhaps axonal in physiology can appear demyelinating, and that can create confusion and overdiagnosis. I think the other aspect of it, too, is maybe not adhering to guidelines when interpreting our electrophysiology. So there are EAN/PNS criteria, as we've talked about in previous episodes, that really outline the criteria for what is weakly supportive, what is strongly supportive of demyelination, and really adhering to those criteria are important when making these diagnoses.

I think the other aspect of it, too, not so much from an electrophysiology standpoint but from a clinical standpoint, is that there's a lot of overlap, especially early on in the disease course with CIDP and other disorders. So, for example, motor-predominant disorders like, for example, lower motor neuron-predominant variants of ALS, even perhaps multifocal motor neuropathy, if we're thinking of a multifocal presentation of CIDP, there can be some overlap, which can lead to some misdiagnoses as well.

So I think it's really important when you have a case of potential CIDP that, number one, you're performing electrophysiology to the utmost technical standards, that you're really using the criteria to make the diagnosis of CIDP. And in those cases where there may be a question based on the history, the exam, really broadening your differential, really thinking about maybe using other ancillary studies like CSF analysis, like maybe nerve ultrasound in centers where that's available, and other lab work, other imaging studies can come in very handy.

Dr. Allen:
Thanks, Nick. That's a terrific summary of some of the diagnostic challenges in CIDP.

We've found that about half of people with CIDP, that carry a diagnosis of CIDP, actually don't have it. And in some settings, it may be even higher than that.

When we're thinking about CIDP, the phenotype of CIDP, the umbrella of CIDP where there's typical and variants, do you think there's some of those variants that are more challenging to diagnose than others or more commonly misdiagnosed than others?

Dr. Silvestri:
Absolutely. I think, in my experience, probably the biggest overdiagnosis is when it comes to sensory CIDP. And I think in these cases, for the most part, what I'm seeing are patients are diagnosed with sensory CIDP, but in point of fact they have more of a distal symmetric axonal polyneuropathy when you really go back and examine people and do the electrophysiology.

I would say the other variant that I've seen frequently misdiagnosed is the motor variant. And again, those tend to be patients, in my experience, that end up having something like motor neuron disease.

Dr. Allen:
Yeah, I agree. And the distal variant, too, can be really, really tricky. So these aren't patients with numbness and tingling and pain that progress slowly over years; it's really a different, different pattern with ataxia and gait instability and hand problems as well. So thinking about the differential for those different variants, I think, is really, really, really helpful before landing on that diagnosis, especially considering all the treatment implications.

Well, this has been a great brief discussion. I think our time is up. Thanks for listening.

Announcer:
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