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Decoding CIDP: Exploring Common and Atypical Variants

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. Can you help us understand the clinical features of CIDP and its variants?

Dr. Silvestri:

Yeah, of course, Jeff. So most cases of CIDP are what we refer to as typical CIDP. In these cases, people will come to us with progressive weakness, numbness, imbalance that's fairly symmetrical and involves both upper and lower extremities. And by definition, CIDP has to have progression at least 8 weeks or longer.

There are many variants of CIDP. Again, the most common being the typical variant that I just mentioned. But such variants include a motor-only phenotype, where it's just motor nerves that are involved, so patients have weakness but no sensory symptoms. There's the converse. There's a sensory variant where patients have sensory loss, which can present as numbness, ataxia, usually a combination of both but without frank weakness on examination. There's a multifocal phenotype which, as it suggests, is not symmetrical. It typically involves motor and sensory involvement in upper and lower extremities but can be variable and can follow main nerve patterns. And then finally, there is a distal phenotype which, as it suggests, the majority of the symptoms are predominantly distal in the hands and in the feet. Again, both of a motor and sensory phenotype.

So those are the typical ways that patients will present. Again, the more common form being the typical form with the combination of weakness, sensory loss, and, again, that sensory loss presenting as typically numbness or ataxia.

Jeff, anything to add? Anything I missed?

Dr. Allen:

Yeah, that's a great summary. One great resource that one can use in order to help understand these definitions are the EAN/EFNS/ENS guidelines that we published in 2021, and those kind of go over these specific definitions of typical and its variants.

And as you highlighted, the main variants are the sensory phenotype, the motor, the distal, and the multifocal. One of the things that the guidelines highlight is that when you are dealing with these variants, they should otherwise have features of typical CIDP. So in other words, the motor variant should have both proximal and distal relatively symmetric weakness, the sensory variant relatively symmetric proximal and distal numbness, and then the multifocal, several areas of both numbness and weakness. And then the distal, it's numbness and weakness usually below the knee and in the hands. But otherwise, the progression should look the same and the reflexes should look the same, generally depressed except maybe not in areas that are clinically not affected.

But that pattern should really be similar. We know those patients that have those variants, those are often the ones that are hardest to diagnose, especially the distal ones in the sensory ones. So seeing those other features that are in common with the typical CIDP are really, really important.

Anything else that –

Dr. Silvestri:

No, I think that's a great point. I mean, I think that really paying attention to the guidelines and holding those guidelines stringently are important, especially as you point out, not so much in the case of typical CIDP, though it's very important, but in those variants, because they're so rare, I think it's really key that you look at those other features of CIDP as well just to make sure that you're making the correct diagnosis.

Dr. Allen:

I guess the other comment to make is that one of the reasons we include these patients under this umbrella of CIDP, the syndrome of CIDP, is that we think there's some commonality in either pathobiology or the way that we treat them or other diagnostic data so that we can put them under a similar umbrella and use that diagnosis. Because for the most part, the way we go about treating them is very, very similar.

Dr. Silvestri:

I agree.

Dr. Allen:

Well, this has been a great bite-sized discussion. That's our time. Thanks so much for listening.

Announcer:

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