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Idiopathic Hypersomnia Insights: A Diagnostic Case Study

Announcer:

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Dr. Rodriguez:

Hi, I'm Dr. Alcibiades Rodriguez. I'm a Neurologist and Sleep Specialist at NYU Grossman School of Medicine. Today I'm going to speak about Idiopathic Hypersomnia Insights: A Diagnostic Case Study.

We have this 26-year-old woman present with excessive daytime sleepiness for over 10 years. She has struggled all this time getting out of bed, using multiple alarms, and may be late for work sometimes, despite sleeping 9 to 10 hours. With the weekend, she is sleeping 12 to 16 hours, and yet she does not feel rested. Every morning, she feels slow and groggy for up to 40 minutes after waking up, and the naps that she takes during the day, either short or long, are unrefreshing. She also has a history of headaches and a couple of syncopal episodes on history. Currently takes no medications. She was thought to be depressed in the past, it was suggested to take medication, but she declined to use them. At some point, she had a home sleep test that showed no sleep-disordered breathing, normal. Alright? She reports no snoring or restless sleep. Physical examination is normal with a BMI of 23. The Epworth Sleepiness Scale is 16. There is no history of cataplexy and no other past medical history, such as head trauma or CNS infections.

So, she presents to you, and you decide to test her. You do an actigraph for a couple of weeks before coming to the lab for testing and show an estimated daily sleep time of 11 to 14 hours. And the PSG that you do then shows a total sleep time of 510 minutes with no sleep-disordered breathing or periodic limb movements of sleep. The sleep efficiency is high at 98% with 25% of slow wave sleep with no SOREMPs. Right? The MSLT showed mean sleep latency of 6.5 minutes with no SOREMPs. Due to this, you diagnose her with idiopathic hypersomnia. She exactly fits the criteria by the American Academy of Sleep Medicine of idiopathic hypersomnia: excessive daytime sleepiness well documented, no cataplexy, right, you did the testing with a PSG showing no SDB, or sleep-disordered breathing, PLMs, right, she has slept 510 minutes during that time, the MSLT shows a mean sleep latency of 6.5 minutes with no SOREMPs, so clinically she qualifies for idiopathic hypersomnia. It's not the history of insufficient sleep or the causes of her sleepiness. Even though you even went further and documented for 2 weeks on actigraph the patient is sleeping over 11 hours. So that shows that she has idiopathic hypersomnia, so she fits the criteria.

Which other supported criteria you may have for this patient? Well, severe and prolonged sleep inertia, she said it takes 40 minutes to get her going in the morning. Some episodes of half awake, half asleep, sleep drunkenness it's called; unrefreshing naps, right, autonomic instability, she mentions some headaches, a couple of times she passed out, they may have also Raynaud's phenomenon, right? Memory and attention difficulties may be due to the sleepiness, right? Depressive symptoms may have these patients, 15 to 25%, in her case, she has very high sleep efficiency too at 98%. So all those are supportive criteria of IH.

This graphic is very important. We need to take into account the differential diagnosis of excessive daytime sleepiness. Right? The number one cause of excessive daytime sleepiness in the world is insufficient sleep. So, I need to be taking it into account; if sleep-disordered breathing especially is moderate to severe; narcolepsy type 1/type 2; idiopathic hypersomnia; other CNS hypersomnias due

to infection, stroke, any brain insult technically, can provoke hypersomnia; traumatic brain injury; medications, legal or illegal, can cause sleepiness; circadian-rhythm sleep disorders; any medical condition; and controversial but worth to mention, depression.

What else can help you to elucidate the diagnosis? Well, excessive daytime sleepiness with no cataplexy. That may be a hint. The sleep drunkenness seems to be more common in IH diagnosed as CNS hypersomnia. So maybe a clue. As I mentioned before, headaches and autonomic features, tension type, or migraine, syncope, as patient had, the unrefreshing naps, long unrefreshing sleep, all those things could be a clue. No other sleep disorder can explain these features. And if you think that the patient has this and testing is not enough, you can repeat the testing, because the clinical correlation is the most important issue.

Thank you, and hopefully that was helpful for you.

Announcer:

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