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Case: Clinical Features of Narcolepsy

## Announcer:

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## Dr. Krieger:

Thank you so much for joining us today. My name is Ana Krieger. I'm a Professor of Clinical Medicine at Weill Cornell Medical College and the Chief of the Division of Sleep Neurology. We'll be discussing today a clinical case with features of narcolepsy.

So, this is a very interesting case. A 42-year-old man with an underlying diagnosis of ADD and Bipolar disease that presented to his neurologist for an evaluation. He had been previously treated with stimulant medications and antidepressants over the years without a significant improvement. Currently, he had been taking methylphenidate up to 148 mg, and still describes significant daytime sleepiness. The neurologist found that his condition was unusual and referred him for a sleep evaluation.

During his sleep consultation, the patient described having significant daytime sleepiness since middle school, unable to concentrate, falling asleep in class, and failing his exams. He would fall asleep as well inappropriately, missing his stops on the buses and subways, and would drive very fast in order to avoid falling asleep while driving. He would drink large amounts of caffeine to offset sleepiness without a significant improvement. When going to bed, he had an immediate sleep onset. And on his score on the Epworth Sleepiness Scale was 21 out of 24.

As an adult, he kept a regular sleep schedule, going to bed between 11pm and getting up at 6:30 in the morning, had very vivid dreams often at night, described a sleep paralysis when waking up, and also endorsed visual hallucinations predominantly at sleep onset, often with repetitive content. And those episodes would also happen when he was napping. When laughing very hard, he would describe episodes of decreasing muscle tone on his neck and feeling that he could not participate in a lot of activities that involved a lot of emotion or anger. He would feel that he may have episodes where he would have to sit down. He would nap daily for up to 3 hours, sometimes multiple times a day.

Over the years, as we discussed, he received multiple diagnoses including ADD and Bipolar disease, cognitive impairment due to poor memory. He had one episode of significant depression at age 28 that required hospitalization. And since then, he had been treated with multiple antidepressants without a clinical improvement. The patient had been avoiding alcohol as he felt it exacerbated his daytime sleepiness.

He had a normal physical exam, a normal BMI, normal neurological exam as well. Therefore, an overnight sleep study was requested.

Before doing the overnight study, the patient was advised to wean off the stimulants because the goal of the study was to evaluate for narcolepsy. So, overnight polysomnography followed by a Multiple Sleep Latency Test was performed. The PSG was normal, normal AHI, although there was an evidence of some restless legs before falling asleep. Multiple Sleep Test was abnormal with an average latency of 1.4 minutes in the presence of 4 sleep onset REM periods.





So clinically, after discussion of the result, the patient was first informed about his new diagnosis, and we discussed safety measures and discussed the expectations of the disease. We started him on a wake-promoting agent. The dose was escalated on modafinil up to 600 mg without a significant clinical improvement. Therefore, we decided to move on to another agent, starting him on oxybate, which the dose was escalated over a period of 4 weeks to the max dose of 4.5 grams twice a night. The patient had very significant clinical improvement, with a reduction on his Epworth scale to 7 at 3 weeks, and to 3, which is very normal, at 6 weeks post treatment. And subsequent psychiatric evaluation including neuropsychiatric testing was normal.

So, some of the important key points of this study for us to remember, if patient has significant daytime sleepiness that had not been fully explained by other diagnosis, had sleep paralysis, vivid dreams, cataplexy also had comorbid restless legs. So, it's important for us to realize that sometimes a diagnosis of narcolepsy may be delayed and may be masked by other underlying potential psychiatric misdiagnosis. So, we hope that you use this information to really monitor your patients and help diagnose patients that are still undiagnosed.

Thank you so much for your attention.

#### Announcer:

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