NREM Sleep Parasomnia Associated with Chiari I Malformation

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CASE REPORTS

Parasomnias are common sleep disorders in children, and most cases resolve naturally by adolescence.¹ They represent arousal disorders beginning in NREM sleep and are generally non-concerning in children. The diagnosis can usually be made by clinical assessment, and testing with polysomnography is not routinely indicated.² However, in certain cases with

atypical features, polysomnography and more extensive neurologic evaluation are medically indicated.

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REPORT OF CASE

A 14-year-old male was referred for evaluation of increasingly frequent nocturnal awakenings with somnambulism since 2 years of age. Multiple episodes occurring predominantly during the first third of the night, characterized by awakening with confusion and anxiety were reported. Somnambulism was often observed, during which he seemed incoherent but exhibited no violent, aggressive behavior or self-injury. There was no recall of these episodes the following day and no associated seizure-like activity. There was no snoring or witnessed apnea; however, restlessness was reported. There were no symptoms of restless legs syndrome or nightmares, and average sleep duration of 9-11 hours was reported. He was tired and had daytime somnolence but no other features of narcolepsy.

He had good academic performance with no substance abuse. Family history was notable for somnambulism in mother and sleep apnea in father. Past history significant for viral meningitis at age 3 months without residual deficits, strabismus surgery at age 7 years, and adenotonsillectomy at age 10 for presumed obstructive sleep apnea causing sleep disruption. He was diagnosed with Asperger syndrome at age 9 because of anxiety and difficult peer relations. He took no prescription medications.

On examination, his height was at the 70th percentile, weight at 50th percentile, and BMI at 45th percentile. There were no craniofacial risk factors for sleep apnea. His neurologic and physi-

cal examinations were normal apart from a high-pitched voice and mild clumsiness.

He was evaluated with polysomnography and noted to have an apnea hypopnea index of 67/h (**Figure 1**). During the first half of polysomnography, 4 confusional arousals occurred out of slow wave sleep (**Figure 2**). There was no evidence of abnormal EMG activity or behavior disturbances during REM sleep.

Cine-magnetic resonance imaging study performed to determine the location of possible airway obstruction showed a Chiari type 1 malformation, with severe protrusion of the cerebellar tonsils extending 19 mm below the foramen magnum, as well as a moderate syrinx of the cervical spinal cord extending from the cervical 2nd to 7th vertebra (**Figure 1**). Two weeks following posterior fossa craniectomy and decompression surgery, complete resolution of the parasomnia occurred. Polysomnography performed 8 weeks later revealed improvement in sleep apnea. Of particular interest was clinical improvement in social skills, coordination and resolution of the high-pitched voice within a month after surgery. Three years following surgery, he has had no further parasomnias.

DISCUSSION

Chiari malformation (CM) consists of inferior displacement of parts of the cerebellar vermis, tonsils, pons, and medulla. Syringomyelia and/or syringobulbia have been associated with CM. Since the original descriptions of the malformation, knowledge of the pathophysiology has evolved; and improved neurodiagnostic techniques enable more accurate detection and correlation of clinical symptoms.³

Clinical manifestations are usually due to impaired cerebrospinal fluid dynamics causing increased intracranial pressure or brainstem dysfunction leading to specific neurologic deficits. Of interest, a study has reported speech difficulties in CM, in which patients' speech quality was altered or they were unable to modulate their voice.⁴

Figure 1—Summary of polysomnographic and magnetic resonance imaging data

TST, Total sleep time; SE, sleep efficiency; PLMS, periodic limb movements in sleep; CAP, Cyclic alternating pattern; AHI, Apnea hypopnea index; OAI, Obstructive apnea index; CAI, central apnea index; HI, Hypopnea index; SpO₂, oxygen saturation.

Figure 2—Polysomnographic demonstration of confusional arousal occurring out of slow wave sleep in the patient

Due to involvement of the brainstem and lower cranial nerves, a strong association between CM and sleep disordered breathing is not surprising. In a recent study involving 46 patients, sleep apnea was present in 70% with CM type I and 50% with CM type II.⁵ Improvement in sleep disordered breathing following decompression surgery is reported.⁶

Although our patient had clinical features that have been associated with CM, the diagnosis was made serendipitously. The presenting symptoms of increasingly frequent confusional arousals and somnambulism have not been previously associated with CM. Polysomnography confirmed that all episodes arose out of NREM sleep. Following surgical decompression, the parasomnia resolved, sleep disordered breathing improved, and vocal cord dysfunction and socialization improved. The

likely mechanism underlying the parasomnia was instability of slow wave sleep triggered by respiratory disturbances.⁷ Surgical decompression improved the apnea hypopnea index with secondary reduction in sleep state instability and the parasomnia. The cerebellum has been implicated in higher cognitive function due to neural pathways connecting the frontotemporal cortex with cerebellum.⁸ Dysfunction of these pathways by elevated intracranial pressure could also be an explanation for the patient's impairment in socialization and parasomnia. Relief of pressure on cranial nerve nuclei likely explains the improved vocal cord function post surgery.

Our case underscores the importance of polysomnography in evaluating patients with parasomnia particularly when the symptoms do not follow the expected natural history of resolu-

tion with age. This case also illustrates the importance of considering an underlying CM when there are multiple confusing symptoms possibly explained by brainstem dysfunction, for which typical risk factors may not be obvious.

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