0820

SLEEP AND OCULOPHARYNGEAL MUSCULAR DYSTROPHY: DISEASE PROGRESSION AFFECTING VENTILATORY NEEDS AND TREATMENT OF SLEEP-DISORDERED BREATHING

Susan Muraida[1], Nicholas Cutrufello[2], Melissa Begay[3], Madeleine Grigg-Damberger[1], Joseph Kern[4]
University of New Mexico Sleep Disorders Center [1] Sleep Center at New Mexico Veterans Affairs Medical Center [2] New Mexico Veterans Affairs Medical Center [3] New Mexico [4]

Introduction: Oculopharyngeal muscular dystrophy (OPMD) is an autosomal-dominant, late-onset, and progressive disease characterized by ptosis and dysphagia, sometimes proximal limb weakness and gait abnormalities. It often presents in patients in their 50s. The progressive functional decline of the pharyngeal muscles results in feeding difficulties and aspiration; however, patients may also have risk of nocturnal hypoventilation and sleep apnea, complicated by variable airway obstruction and compliance.
Report of Cases: Using retrospective chart review, we identified patients with a known diagnosis of OPMD treated at the Raymond G. Murphy VA Sleep Center. We present a case where OPMD progression necessitated increased ventilatory support and affected positive airway pressure (PAP) compliance. A 58-year-old male with OPMD, DMT2, depression, and memory impairment underwent home sleep apnea testing showing severe OSA (REI 32.7, SpO2 nadir 72%). He started Auto-PAP 6-16 cwp and presented to discuss issues tolerating PAP. Pressures were lowered, but he continued to require maximum pressures without increased utilization. An in-lab CPAP titration showed treatment-emergent centrals but did not find optimal pressures due to limited sleep time. Having failed CPAP, he returned for an ASV titration which controlled his apnea in lateral position. Patient was switched to auto ASV to increase efficacy and comfort. Two months later he discontinued ASV due to frustration with disease progression and feeling unable to breathe deeply with the machine. Nocturnal oxygen at 1L was ordered while he awaited Neurology consult for OPMD. Later, concerned about progressive dyspnea, he resumed ASV, now with 3L O2 bleed. Given suspicion of hypoventilation (bicarbs 27-30), and that ASV could not adjust to his continually changing airway tone with his OPMD, he was switched to iVAPS. This resulted in good control of his sleep apnea, tidal volumes and minute ventilation. However, he reported pressures felt too high, returned to ASV for a period, then discontinued PAP altogether.
Conclusion: Patients with OPMD and sleep apnea require close follow-up as their disease progression may affect their ventilatory support needs. These patients may require more complex PAP modalities, such as AVAPs, and routine PFTs to help determine timing of ENT involvement for surgical airway planning.
Support (If Any):

0821

NARCOLEPSY AFTER WEST NILE VIRUS MENINGOENCEPHALITIS INFECTION

Kishan Tarpara[1], Safia Khan[1]
UT Southwestern [1]

Introduction: Narcolepsy is a sleep disorder characterized by hypersomnia and inappropriate intrusion of REM sleep into wakefulness. Etiology is heterogeneous, and is broadly classified into Narcolepsy Type 1 and Type 2. There is growing evidence that the pathogenesis in many cases have a post-infectious autoimmune basis. We present the case of a patient seen in our sleep clinic who was eventually diagnosed with Narcolepsy type 2 following West Nile Virus (WNV) Meningoencephalitis.
Report of Cases: · The patient is a 44 year old Hispanic woman who had no prior sleep difficulties before 2013, when she was diagnosed with WNV meningitis. Lumbar puncture results from the time of infection are not available, as this evaluation occurred in the Dominican Republic. Subsequent lumbar punctures have shown the presence of anti-WNV IgG antibodies in her CSF. MRI and MRA brain were unremarkable.
· She also has a history of migraine, fibromyalgia, depression, and adrenal insufficiency.
· She presented with sleep complaints, including sleep onset insomnia, symptoms of sleep
disordered breathing, and excessive daytime sleepiness.
· She underwent home sleep apnea test in 12/2020 which revealed mild obstructive sleep apnea. Overall AHI was 7.9/hour without significant hypoxemia. She was subsequently started on CPAP therapy with good adherence, but she continued to have significant daytime sleepiness, notably with an Epworth Sleepiness Scale score of 21/24.
· She subsequently underwent overnight polysomnogram and multiple sleep latency test in our sleep laboratory.
· Overnight PSG confirmed mild obstructive sleep apnea (AHI 12.0) along with short REM latency (10.5 minutes). Next day MSLT showed mean sleep latency of 2.3 minutes, for 5 attended naps, with 2 SOREMPs, which was diagnostic of Narcolepsy type 2.
Conclusion: This case demonstrates the onset of narcolepsy after WNV meningoencephalitis.
Support (If Any):

0822

CAFFEINE AS A TREATMENT OPTION FOR PRIMARY CENTRAL SLEEP APNEA OF INFANCY IN TERM INFANTS

Amit Shah[1], Ajay Kasi[1], Samar Shah[1], Fran Martinez[1], Roberta Leu[1]
Emory University School of Medicine/Children's Healthcare of Atlanta [1]

Introduction: Primary central sleep apnea of infancy tends to improve over weeks with supportive care. No established treatments exist; however, infants with this condition remain at risk from sequelae of intermittent hypoxemia. We present a term infant with primary central sleep apnea of infancy treated with caffeine citrate resulting in clinical and polysomnographic improvement.
Report of Cases: A 7-day-old male infant born at 37 weeks gestation (gestational age confirmed by early first trimester prenatal ultrasound) was hospitalized following an episode of hypotonia and decreased responsiveness. Infectious studies, chest radiograph, and echocardiogram were normal. During the hospitalization, oxygen desaturations during sleep were observed and capillary blood gas during sleep showed a pH of 7.36 and a partial pressure of carbon dioxide of 54 mmHg. Polysomnography on room air showed central sleep apnea [central apnea hypopnea index (AHI) of 58, obstructive AHI of 4, hypoxemia, hypoventilation with transcutaneous carbon dioxide greater than 50 mmHg for 89% of sleep time, and periodic breathing for 21.7% of sleep time]. Brain MRI and paired-like homeobox2B (PHOX2B) genetic testing were normal. A trial of caffeine citrate was initiated with