

Sleep and Breathing, continued

report an unusual occurrence of MG presenting for the first time as Myasthenic Crisis in the post partum period.

CASE PRESENTATION: Our patient is a 21-year-old African American Female G3P2103 who was successfully extubated in the operating room following emergent Caesarian section (for cord prolapse) under general endotracheal anesthesia and succinylcholine. Two hours post-operatively while in the recovery room the patient was noted to have a blood pressure of 180/120 with tonic-clonic shaking movements. She was reintubated for airway protection after receiving boluses of succinylcholine and magnesium sulfate. She was subsequently placed on mechanical ventilation and started on a magnesium drip for eclampsia and empiric broad spectrum antibiotics including gentamicin. Soon after, all sedation was stopped and the magnesium drip was discontinued. Five hours following reintubation, the patient was awake, alert and following commands while continuing to require ventilator support. Her vital signs remained stable. Neurological exam was performed and was remarkable for bilateral ophthalmoplegia. The pupils were bilaterally equal and reactive to light but she was unable to track objects. The oculocephalic (Doll's eyes) reflex was also absent. Surprisingly, she was able to visually track objects by lifting and turning her head. Her deep tendon reflexes were brisk and symmetrical with normal motor strength and an intact sensory system. A Computed Tomography (CT) scan of the head was normal. Our findings suggested bilateral ocular myopathy in the setting of acute respiratory failure. Our differential diagnoses included; Myasthenia Gravis, Botulism, Tick paralysis, Lambert Eaton Syndrome, Organophosphate poisoning and Guillain-Barre Syndrome. Electromyography with repetitive nerve stimulation revealed decremental motor response consistent with Myasthenia Gravis. This was later confirmed by positive serum acetylcholine receptor antibodies. Our patient was started on plasmapheresis. Her weaning parameters started improving after the second plasma exchange and she was successfully extubated after the third session. A routine CT of the chest demonstrated an age related excess of thymus tissue.

DISCUSSIONS: Currently with the advent of therapy the incidence of MC has diminished. This, however, does not pertain to those Myasthenia patients who initially present in crisis, as seen in our case. MC not only requires timely recognition but also the identification of precipitating factors. Such factors noted in our patient included the stresses of pregnancy and surgery, the use of medications such as a neuromuscular junction blocker (succinylcholine), magnesium sulfate and an aminoglycoside (Gentamicin). The current treatment modalities for MC include plasmapheresis and IV immunoglobulin, while trials with steroids are reserved for non-responders. Anticholinesterase drugs and immunomodulators are the cornerstone for chronic and maintenance therapy.

CONCLUSION: Neuromuscular causes should be considered in all young patients presenting with acute respiratory failure, with a special consideration for MC in those within the specific sex, age group, and precipitating factors.

REFERENCE:

1. Lacomis D., Myasthenic crisis. *Neurocritical Care*. 2005;3(3) 189-94.

DISCLOSURE: Peggy Rahal, None.

A 66-YEAR-OLD WOMAN WITH OBSTRUCTIVE SLEEP APNEA AND HYPOXEMIA

Nehal P. Thakkar MD^{*} Oren P. Schaefer MD, FCCP University of Massachusetts, Worcester, MA

INTRODUCTION: Isolated drainage of the superior vena cava (SVC) into the left atrium is an extremely rare finding, with only 15 cases reported in the literature. The majority of reports describe this anomaly in adolescents; the number of adults diagnosed with this congenital abnormality is very few. We report a 66-year-old female referred for obstructive sleep apnea who was found to have such an anomaly.

CASE PRESENTATION: The patient is an obese 66-year-old female with a past history of breast cancer, treated with left mastectomy, who was seen for evaluation of obstructive sleep apnea syndrome (OSAS). A polysomnogram showed sleep apnea with desaturation well out of proportion to the degree of apnea. Complaints included restless sleep with frequent awakenings, bifrontal headaches, and dyspnea with exertion. Pulmonary function tests demonstrated normal spirometry, normal diffusing capacity, and lung volumes significant for a reduced expiratory reserve volume. Physical examination was remarkable for weight of 281 pounds, and room air oxygen saturation of 91%. There was no jugular venous distension, heart sounds were regular, no murmurs were auscultated,

and the chest was clear. There was no cyanosis. A chest x-ray was unremarkable. An arterial blood gas showed a pH of 7.40, pCO₂ of 42, and a pO₂ of 62 on room air. Given the degree of hypoxia, the patient underwent further testing. A chest CT scan, did not show any parenchymal disease, but did show a persistent left sided SVC. A 100% oxygen shunt study found a shunt of 26%. A nuclear perfusion shunt scan with macroaggregated albumin revealed marked tracer uptake in extra pulmonary tissue when injected into the right arm - 92% of the tracer was present outside of the lungs. A transthoracic bubble echocardiogram demonstrated contrast entering the left atrium directly after injection in the right antecubital vein. A transesophageal echocardiogram confirmed a left sided SVC entering an enlarged coronary sinus, and a right-sided SVC entering the left atrium. Injection of contrast in the right arm confirmed communication of the right SVC with the left atrium. Contrast injection in the left arm showed communication of the left SVC with the coronary sinus.

DISCUSSIONS: This report details a 66-year-old female who was found to have a congenital large vessel right to left shunt with drainage of the right SVC into her left atrium. Only 15 similar cases have been reported. This patient appears to be the oldest reported patient with this rare abnormality. Embryologists theorize that malposition of the right horn of the sinus venosus in a leftward and cephalic direction, with subsequent connection of the dominant SVC to the left atrium, rather than the right atrium, underlies this anomaly. Of clinical importance, the patient had been advised to avoid all intravenous infusions and utilization of the veins of her left arm given her mastectomy on that side. Given the above findings, the patient was advised to avoid infusion into her right arm to decrease the risk of systemic embolization in the presence of the right to left shunt.

CONCLUSION: This case highlights the fact that wakeful hypoxemia is not consistent with pure OSAS and should prompt the clinician to explore this further. Evaluation in this case pointed to a right to left shunt and the diagnosis of an extremely rare congenital malformation, undetected well into adulthood.

REFERENCES:

1. Rosenkranz S, et.al. Anomalous drainage of the right superior vena cava into the left atrium in a 61-year-old woman. *Int J Cardiol* 1998; 64:285-291.
2. Ezekowitz, MD, et.al. Isolated drainage of the superior vena cava into the left atrium in a 52-year-old man. *Circulation* 1978; 58:751-756.

DISCLOSURE: Nehal Thakkar, None.

INTRACRANIAL HYPERTENSION CAUSED BY OBSTRUCTIVE SLEEP APNEA AND OBESITY HYPOVENTILATION SYNDROME

Christine H. Won MD, MS^{*} Carl Kirsch MD Stanford University Medical Center, Stanford, CA

INTRODUCTION: This report describes a case of papilledema and intracranial hypertension caused by obstructive sleep apnea (OSA) and obesity-hypoventilation syndrome (OHVS).

CASE PRESENTATION: A 23-year-old morbidly obese man presented to the emergency room with 4 months of headaches and progressive bilateral constrictive peripheral visual field loss. Fundoscopic exam revealed moderate bilateral papilledema. Intraocular pressure in each eye measured 23 millimeters of mercury (mmHg). A lumbar puncture showed an elevated opening pressure of 38 mmHg. Cerebral spinal fluid (CSF) studies and magnetic resonance imaging of the brain were normal. The patient was diagnosed with idiopathic intracranial hypertension (IIH). He was started on acetazolamide, and underwent 5 therapeutic lumbar punctures to remove a total of 250 milliliters of CSF. The patient showed no clinical improvement and he was referred for a ventriculo-peritoneal CSF shunt. During his hospital stay, the patient was noted to be chronically hypoxic with a pulse oximetry of 88%. His hematocrit was elevated to 61%. His serum bicarbonate level was 28 millimoles per liter. An arterial blood gas (ABG) while breathing air showed a pH of 7.29, pCO₂ 54 mmHg, and pO₂ 58 mmHg. The patient revealed a long history of loud snoring and excessive daytime somnolence. A nocturnal polysomnography confirmed severe OSA, with 118 apneic and hypopneic events per hour, and a pulse oximetry nadir of 55%. The patient was started on nocturnal noninvasive mechanical ventilation with supplemental oxygen for OSA and OHVS. Two days later, the patient reported improvement in his vision and headaches. His ABG improved to pH 7.31, pCO₂ 43 mmHg, and pO₂ 80 mmHg. The lumbar puncture opening pressure