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Locked in syndrome: A case report

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Synopsis: We report a case of a 53-year-old male, newly diagnosed with hypertension, presented with left-sided body weakness and numbness and decreased verbal output. Over 10 days, patient had been experiencing intermittent rotatory dizziness, no history of trauma nor loss of consciousness. After presentation, patient became quadriplegic, anarthric and presented an initial period of coma, requiring intubation and ventilatory assistance. He was started on a neuroprotectant, low molecular weight heparin, antiplatelet, antidyslipidemiac agent, oral antihypertensive agent on day of admission. Early intensive rehabilitation and family counselling were done. While admitted, patient developed ventilator acquired pneumonia and eventually expired on his 30th day of hospitalization.

Clinical Presentation: Patient is M.B. 53 year old male, Filipino, right handed, presented with left-sided body weakness and numbness. 10 days prior to admission, patient experienced intermittent rotatory dizziness with slurring of speech relieved by rest. One day prior to admission, with recurrence of dizziness, still with slurred speech, he sought consult in a private Clinic with BP of 190/100, diagnosed with BPPV and Hypertension Stage II, given medications. 5 hours prior to admission, patient complained of left-sided body numbness with weakness and was noted to have decreased verbal output but can comprehend and follow some commands. He sought consult and subsequently admitted. Patient is recently diagnosed with Hypertension Stage II, prostatic enlargement on Tamsulosin 200mcg, 1 tab OD. Patient's father has hypertension, siblings has Type 2 DM and heart disease on maternal side. He was nonsmoker, occasional alcoholic beverage drinker, denies illicit drug use. He was a retired seaman residing in USA, and a volunteer employee at Home of Aged in USA.

Physical Findings: At time of examination, patient was awake, conscious, coherent , not in distress with vital signs as follows: BP 170/90 (MAP 117 mmHg), HR 62 beats per minute, RR 20 cycles per minute, afebrile at 37degrees with 02 saturation of 98%. He was average built male, weigh 75kg, BMI 27.5 kg/m2. On neurological examination, GCS 11 (E4V2M5), NIHS Score of 19, 2-3 mm pupils sluggishly reactive to light, AVR ratio 2:3, no signs of hypertensive retinopathy, preferential gaze to right with full extraocular muscle movement, left central facial palsy, bilateral weak gag reflex, cannot shrug left shoulder because of left sided weakness, left hemiplegia of 2/5 upper and 1/5 lower extrimities. With the same degree of painful stimulation, there is a delay in the response over the left upper and lower extemities.

Laboratory Work-up: CBC, electrolytes, kidney and liver functions tests, chest xray, urinalysis and KUB with Prostate Ultrasounds were normal. Plain Cranial CT showed chronic infarct, right subinsular region with mild microvascular ischemic disease and atherosclerotic intracranial vessel disease. Cranial MRI showed acute to subacute infarct at anterior two-thirds of the pons, chronic infarct at periphery of the left pons and right lentiform nucleus; atherosclerrotic internal carotid arteries; occlusion or very slow flow in the distal vertebral arteries and proximal basilar artery. Transcranial doppler ultrasound was normal. Carotid Duplex Scan showed <50% stenosis at right common carotid artery, 50-59% stenosis right internal carotid artery, <50% (1-15%) stenosis left internal carotid artery, indicative of a more distal occlusive in the posterior circulation. Echocardiography showed mild to moderate aortic regurgitation and Grade 1 left ventricular diastolic dysfunction.

Diagnosis:Locked-In Syndrome; Hypertension Stage II; Ventilator Acquired Pneumonia

Treatment:Medical management started on neuroprotection, antiplatelet, low molecular weight heparin, maintaining an airway and adequate oxygenation via mechanical ventilator, tracheostomy and gastrostomy were done, early intensive rehabilitation, and family counselling.

Outcome :While admitted, patient developed ventilatory acquired pneumonia as caused of demise of patient.

Significance: Locked In Syndrome (LIS) is a rare neurological condition characterized by complete paralysis of voluntary muscles in all parts of the body except control of eye movement, preserved cognitive functioning and a primary code of communication that uses vertical eye movements or blinking. This condition leaves the individual completely mute and paralyzed. Prevalence is unknown. Their only means of communication is by blinking or vertical eye movements because of sparing of the midbrain tectum, which allows communication.

Recommendations:Locked-in syndrome present as a great challenge to internists, hence, thorough investigation is needed in arriving at diagnosis. Internists should do the complete neurological examination and assessment. To our knowledge, there is no known documented incidence in the local setting of Makati. It can be difficult to diagnose and it can be missed if voluntary vertical eye movement is not assessed. With the various new modalities to diagnose LIS, there now exists the possibility to unlock sufferers from this devastating neurological condition.

Biography

Jalellah B Noor has completed her Doctor of Medicine at Far Eastern University - Nicanor Reyes Medical Foundation, Philippines. She has completed her Internal Medicine training at Ospital ng Makati, Makati City, Philippines.

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