Title:

A Case of Hemiplegia Vegetativa Alterna, Paroxysmal Sympathetic Hyperactivity and Ogilvie’s Syndrome: The Role of Central Sympathetic Pathways in their Pathophysiology

Authors and Institution:

Robert SK Chan, Joanna WK Ho, Sharon TM Chan, Peter YM Woo; KM Leung, KY Chan

Department of Neurosurgery, Kwong Wah Hospital

Abstract:

Hemiplegia vegetativa alterna (HVA) is the clinical syndrome of contralateral hemiparesis, hemisensory loss, hemihyperhydrosis and ipsilateral Horner’s syndrome1,2. The term vegetativa alterna denotes that a single brainstem lesion manifests with ipsilateral and contralateral, i.e. crossed, signs of autonomic (“vegetative”) sympathetic nervous system dysfunction. Fewer than five cases have been reported and most were a result of stroke involving the occlusion of posterior cerebral artery (PCA) perforators that supply the anterolateral mesencephalon 1-3.

We describe a 46 year old male who suffered from aneurysmal subarachnoid hemorrhage and exhibited HVA as a consequence of mesencephalic injury. The patient also experienced paroxysmal sympathetic hyperactivity (PSH) and recurrent colonic pseudo-obstruction, known as Ogilvie’s syndrome. PSH is identified as a pathological state of elevated sympathetic activity characterized by episodic tachycardia, hypertension, tachypnea, hyperthermia, diaphoresis and decerebrate dystonia occurring in 10% of severe traumatic brain injury patients4. Although the pathogenesis has yet to be elucidated the excitatory-inhibitory ratio (EIR) disconnection theory postulates that certain brainstem centers are inhibitory in nature and when severely injured, the spinal cord is released from higher regulation leading to sympathetic hyperactivity5. Colonic pseudo-obstruction is a gastro-intestinal emergency with brain injury accounting for 10% of cases6,7. It is understood that sympathetic hyperactivity also plays an important role in its pathophysiology7.

To our knowledge this is the first documented case of HVA, PSH and colonic pseudo-obstruction in the literature. We propose that the three conditions are related to an interruption of central sympathetic pathways and present clinico-radiological evidence to demonstrate that focal lesions of the anterolateral mesencephalon can account for this unique presentation.