Seventh-and-a-Half Syndrome

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Abstract

This is a case of a patient with right internuclear ophthalmoplegia and right peripheral seventh nerve palsy with no other neurologic deficits. Magnetic resonance imaging showed a small localized right hemipons infarct involving facial motor nucleus and facial genu as well as the right medial longitudinal fasciculus. We introduce “Seven-and-a-half syndrome” as a new clinicoradiologic syndrome.

Case Presentation

A 68-year-old white female presented with sudden onset horizontal binocular diplopia and right-sided facial weakness involving the upper and lower face consistent with a severe lower motor neuron seventh nerve palsy. Past medical history was significant for uncontrolled hypertension and cerebral amyloid angiopathy. No history of head trauma or infection. Visual acuity was 20/20 in both eyes. Pupils were equal and reactive with no relative afferent pupillary defect in either eye. Visual field testing was normal bilaterally. Patient had no ptosis. Motility showed deficit in adduction of the right eye with a dissociated horizontal abducting nystagmus in the left eye. Slit lamp exam showed punctate epithelial erosions on the right cornea consistent with exposure keratopathy secondary to the seventh nerve palsy. The remainder of the motor and sensory examinations was normal. Magnetic Resonance Imaging (MRI) of the brain showed an evolving 3mm acute lacunar infarction in the posterior right hemipons, in the right facial colliculus involving the facial motor nucleus and facial genu as well as the right medial longitudinal fasciculus (Figure 1A and 1B). The patient had a full recovery after a few months.

Several classic clinicoradiologic syndromes have been noted involving the pons. Raymond syndrome [1] involves the ventral pons affecting the CN VI fascicle and the pyramidal tract causing VIth nerve paresis and contralateral hemiparesis. Millard Gubler syndrome [1] includes VIIth nerve paresis, contralateral hemiparesis and an ipsilateral VIIth nerve paresis. This lesion is still ventral but affecting the fascicle of the VIIth nerve as well. The Foville syndrome [1,2] encompasses a VIIth nerve paresis, horizontal conjugate gaze palsy, ipsilateral CN V, VII and VIII palsy and an ipsilateral Horner syndrome. Other pontine syndromes include the unilateral or bilateral Internuclear Ophthalmoplegia (INO) associated with a “wall eye”, exotropia (i.e., WEMINO and WEBINO). A horizontal gaze palsy from the VIIth nerve nucleus in the pons. The combination of the horizontal gaze palsy and an INO has
been referred to as the “one-and-a-half syndrome”. A concomitant CN VIII palsy in this setting has been called the “Eight-and-a-half syndrome”. Our patient had a combination of right INO and right facial nerve palsy with no other neurologic abnormalities (a “seven-and-a-half syndrome”). Clinicians should be aware of the various syndromes that can occur in dorsal and ventral pontine lesions.

References