One and One-half Syndrome With Supranuclear Facial Weakness

Magnetic Resonance Imaging Localization

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Objective: To provide clinicoanatomical correlation for a small pontine tegmental ischemic stroke producing the one and one-half syndrome associated with supranuclear facial weakness.

Design: Case report.

Setting: Tertiary care center.

Patient: A 70-year-old man developed left-sided facial weakness sparing the forehead, a left internuclear ophthalmoplegia, and a complete left horizontal gaze palsy immediately after percutaneous transluminal coronary angioplasty. Magnetic resonance imaging demonstrated a small lesion in the left paramedian aspect of the dorsal pontine tegmentum.

Main Outcome and Results: Electromyographic findings were consistent with supranuclear facial involvement. The patient had nearly complete recovery after 1 year.

Conclusions: To our knowledge, this is the first report of supranuclear facial weakness in association with the one and one-half syndrome. The location of the lesion provides evidence of the existence of corticofugal fibers that extend to the facial nucleus in the dorsal paramedian pontine tegmentum.

Arch Neurol. 1999;56:1509-1511

THE ONE and one-half syndrome consists of a horizontal gaze palsy (“the one”) and an ipsilateral internuclear ophthalmoplegia (“the one-half”). When the eye that is contralateral to the lesion lies in a position of abduction at rest, the syndrome has been called paralytic pontine exotropia. The horizontal gaze palsy results from involvement of either the ipsilateral paramedian pontine reticular formation or the ipsilateral abducens nucleus, and the internuclear ophthalmoplegia results from involvement of the ipsilateral medial longitudinal fasciculus. We describe a man with a small pontine tegmental ischemic stroke, presumed to be embolic, whose clinical findings were limited to the one and one-half syndrome and supranuclear facial weakness. Patients previously described as having facial weakness and the one and one-half syndrome all had a lower motor neuron pattern of facial weakness. To our knowledge, this is the first report of supranuclear facial weakness in association with the one and one-half syndrome, and the location of the lesion was confirmed by magnetic resonance imaging.

A 70 year old man developed horizontal diplopia immediately after percutaneous transluminal coronary angioplasty. There was no history of stroke or visual disturbance. His risk factors for atherosclerosis included hypertension, hyperlipidemia, and obesity.

The results of his general physical and mental status examinations were normal. He had a complete horizontal gaze palsy to the left for voluntary, tracking, and vestibulo-ocular movements. Convergence was impaired, with only minimaladduction of both eyes. A left internuclear ophthalmoplegia was present, with failure of abduction in the left eye and horizontal nystagmus in the abducting right eye. In primary position, the patient had resting exotropia of the right eye and was able to adduct the right eye to midposition when using it for fixation. Vertical eye movements and eyelid function were intact, and the pupils were briskly and symmetrically reactive to light. He had left-sided facial weakness that involved the muscles of the mouth and lower orbicularis oculi, sparing the upper orbicularis
oculi and the muscles of the forehead (a central pattern of weakness), a diminished left corneal reflex, and a slight decrease in pinprick sensation on the left side of the face. The findings of the rest of the neurological examination were normal. All serum laboratory values were unremarkable.

A magnetic resonance imaging scan obtained on hospital day 3 demonstrated a well-defined focus of increased signal character localized to the left paramedian aspect of the dorsal pontine tegmentum, just ventral to the fourth ventricle (Figure 1). The lesion measured $5 \times 3 \times 6\, \text{mm}$. Diffusion-weighted images supported the acuteness of the finding.

The remainder of the patient’s hospital course was uneventful. The results of his examination were unchanged 4 months later.

The patient returned for follow-up after 1 year. He had no complaints and felt that he had returned to normal. On examination, he had conjugate gaze in primary position and a subtle left internuclear ophthalmoplegia. When fatigued, he had a subtle left conjugate horizontal gaze paresis. He had very mild lower facial weakness and normal findings on his sensory examination. A second magnetic resonance imaging scan demonstrated resolution of the abnormality in the pons. Electromyography of the facial muscles showed no evidence of denervation. Blink reflex testing revealed R1 delay on the left, and bilateral R2 delay with stimulation of the left supraorbital nerve, consistent with residual dysfunction of the trigeminal nucleus.

**COMMENT**

Since Leo Tolstoy’s first account of the one and one-half syndrome resulting from a brainstem stroke, the eye movement disorder has been described both as an isolated syndrome and in association with dysphagia, dysarthria, hemiparesis, ataxia, hemisensory loss, and involuntary movements. Causes of the syndrome include multiple sclerosis, hemorrhage, tumors, and ischemic stroke. The anatomical substrate for the syndrome, including involvement of the ipsilateral medial longitudinal fasciculus and either the abducens nucleus or the pontine conjugate lateral gaze center in the dorsal pontine tegmentum adjacent to the fourth ventricle, was first proposed by Fisher and later confirmed by neuropathological and neuroimaging findings. The onset of symptoms in our patient, immediately after an arterial angiographic procedure and without arrhythmia, hypoxia, or hypotension, suggests an embolic source for the stroke. The paramedian pontine tegmentum is irrigated by the long anteromedial group of paramedian pontine perforating branches off the basilar artery.

The left supranuclear facial weakness in our patient is consistent with involvement of corticobulbar fibers that extend to the facial nucleus. The facial nucleus is innervated by fibers that originate in the precentral gyrus. Corticobulbar fibers, including those destined to reach the facial nucleus, descend in the internal capsule in association with the corticospinal tract. Dejerine noted that in the brainstem corticobulbar fibers leave the pyramidal tract as a series of discrete bundles, decussating to reach contralateral cranial nerve nuclei. Among the bundles he identified were a subthalamic bundle that extends to the nucleus of the third cranial nerve (Figure 2, fasth), a peduncular bundle to cranial nuclei III, VI, and XI (Figure 2, fapd), a pontine bundle to cranial nuclei V, X, and XII (Figure 2, fap), and a pontomedullary bundle to the nucleus of the facial nerve that also contributes fibers to the pontine bundle (Figure 2, fabp). This fourth bundle leaves the pyramidal tract at the level of the pontomedullary junction and passes through the pontine tegmentum to reach the facial nucleus. The clinical significance of these bundles is that lesions selectively involving one bundle may produce a restricted pattern of supranuclear weakness. Prior reports have confirmed the presence of supranuclear fibers extending to the facial nucleus in the pons, but the exact course of these fibers through the pons is unknown. Some of the corticobulbar fibers project directly to the facial nucleus, and others connect via pontine reticular interneurons. There are also corticobulbar fibers that descend ipsilaterally in the ventromedial part of the rostral medulla, decussate, and then ascend rostrally to the contralateral facial nucleus. We believe that our patient’s weakness resulted from involvement of the fourth bundle of Dejerine in the pontine tegmentum.
Previous cases of facial weakness with this eye movement disorder describe a lower motor neuron pattern of facial weakness, attributed to involvement of the facial nucleus or fasciculus. It is conceivable that our patient’s upper motor pattern of facial weakness was due to partial damage to the intramedullary fascicles of the seventh nerve, rather than to interruption of corticobulbar fibers to the facial nucleus. However, lesions involving these fascicles typically produce a lower motor neuron pattern of facial weakness. We are unaware of a well-documented example of facial weakness due to a fascicular lesion. The findings of electrodiagnostic testing in our case showed no evidence of a lower motor neuron lesion. One report also describes palatal myoclonus in association with the one and one-half syndrome and facial weakness that is attributed to involvement of the adjacent central tegmental tract.

The one and one-half syndrome with supranuclear facial weakness in our patient with a well-delineated, small ischemic stroke of the paramedian pontine tegmentum represents another variant of this interesting syndrome.

Accepted for publication May 24, 1999.

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REFERENCES