Cheiro-Oral Syndrome with Internuclear Ophthalmoplegia and Cerebellar Ataxia following Midbrain Infarction

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The cheiro-oral syndrome (COS) is a unilateral sensory disturbance that is limited to the hand and the ipsilateral perioral region. Lesions have also been reported in the parietal cortex [1], corona radiata [2], thalamus [3], and pons [4]. Except for one case of a small midbrain hemorrhage [5], midbrain lesions have not been reported. We describe the first case of a small midbrain infarct leading to COS with the rare combination of internuclear ophthalmoplegia and cerebellar ataxia.

A 52-year-old man suddenly developed dizziness with left-sided dysesthesia, at the corner of the mouth, tip of the tongue, and the first 3 fingers on the left hand. He experienced double vision and was admitted to the hospital on the same day. On admission, he was alert and cooperative but his speech was slow and slurred. Isocoria was evident in the pupils and the light reflex was normal. The horizontal gaze to the left showed paresis of adduction of the right eye and coarse horizontal nystagmus of the abducting left eye. Convergence was incomplete. Dysesthesia was evident around the left corner of the mouth, the left side of the tip of the tongue, and the first 3 fingers of the left hand. Muscle power was essentially normal. Deep tendon reflexes were normal and plantar responses were flexor. Slight dysmetria was noted in the limbs on the left. The gait was slightly wide-based and unsteady. A cranial CT revealed no abnormalities.

The adduction paresis of the right eye and nystagmus of the abducting left eye disappeared on the second hospital day. The sensory disturbances and cerebellar signs gradually improved. MRI performed 40 days after onset revealed a small lesion of low intensity on a T1-weighted image, and of high intensity on a T2-weighted image in the right tegmentum of the midbrain between the red nucleus and the cerebral aqueduct.

Fig. 1. MRI on day 40. Small discrete lesion (arrows) of low intensity on a T1-weighted image (A) and of high intensity on a T2-weighted image (B) was disclosed in the right tegmentum of the midbrain between the red nucleus and the cerebral aqueduct.

The combination of a sensory disturbance in the hand and perioral area on the same side is known clinically as the cheiro-oral syndrome. This combination of signs is attributed to lesions in some parts of the brain that include a characteristic somatotopic arrangement of the somatosensory system; the hand and face areas in the somatosensory cerebral cortex, the ventral posterolateral and ventral posteromedial nuclei...
of the thalamus, and the spino- and trigeminothalamic tracts in the brainstem [6]. Indeed, COS has variously been reported to be lesions of the parietal cortex [1], thalamocortical projection [2], thalamus [3] or pons [4]. Although the spinothalamic and ventral trigeminothalamic tracts travel through the midbrain, only in one case was midbrain hemorrhage reported to cause COS [5]. This is the first report of COS to be caused by a midbrain infarct. A small midbrain infarct detectable on MRI possibly involved the ventral trigeminothalamic tract and the medial portion of the medial lemniscus, thus producing COS. In addition, our patient showed transient internuclear ophthalmoplegia and cerebellar ataxia early in his disease. These symptoms were perhaps caused by the transient involvement of the medial longitudinal fasciculus and the decussation of the superior cerebellar peduncle which are closely situated to the midbrain lesion as demonstrated in our case. We consider these combined symptoms to be important in localizing the lesion of COS within the midbrain vs. that caused by lesions in other parts of the pons, thalamus, thalamocortical projection and parietal cortex.

References
