

Radiologic-Clinical Correlation

One-and-a-Half Syndrome Associated with Cheirooral Syndrome

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Lesions that involve the paramedian pontine reticular formation (PPRF) or the sixth-nerve nucleus on one side of the brain stem and also interrupt internuclear fibers of the ipsilateral medial longitudinal fasciculus produce an ipsilateral horizontal gaze palsy combined with an internuclear ophthalmoplegia, so that the only preserved horizontal eye movement is abduction of the contralateral eye (1). This syndrome consists of a complete horizontal gaze palsy when the patient looks toward the side of the lesion, combined with half gaze palsy when the patient looks in the opposite direction, and has been termed the "one-and-a-half syndrome" (2-5).

Clinical History

A 75-year-old woman who had been treated for hypertension for 5 years was admitted to our hospital with the sudden onset of vertigo, diplopia, and dysesthesia of the left side of the mouth and left hand. On admission, her blood pressure was 197/95 mm Hg and her heart rate was 83 beats per minute and regular. She was alert and exhibited no motor disturbance. Neurologic exam-

ination revealed normal motor function and reflexes. The dysesthesia involved the corner of the mouth, tongue, palm, and thumb on the left side. She experienced a burning sensation in the left fingers, especially the thumb, and swelling and heat in the palm. She also reported a burning sensation of the tongue and lip on the left side.

The right eye was fixed at the midline, but the left eye was abducted on forward gaze (paralytic pontine exotropia) (Fig 1A). On right gaze, neither eye could pass the midline (horizontal gaze palsy toward the right side). On left gaze, the left eye was abducted and demonstrated a monocular nystagmus; the right eye was not adducted (medial longitudinal fasciculus syndrome on the right side). Thus, there was complete horizontal gaze palsy when the patient looked toward the right side, combined with half gaze palsy when the patient looked in the opposite direction (one-and-a-half syndrome) (Fig 1A). Vertical eye movements and convergence were not involved.

Hematologic studies, blood chemistries, renal and hepatic function tests, fasting blood sugar, total cholesterol, triglyceride, chest roentgenogram, and electrocardiogram were normal.

Magnetic resonance (MR) 3 days after the onset of the stroke demonstrated three small high-intensity lesions in the dorsal portion of the right pontine tegmentum contacting the floor of the fourth ventricle on proton density- and T2-weighted images (Fig 1B and C). The lesions involved the medial longitudinal fas-

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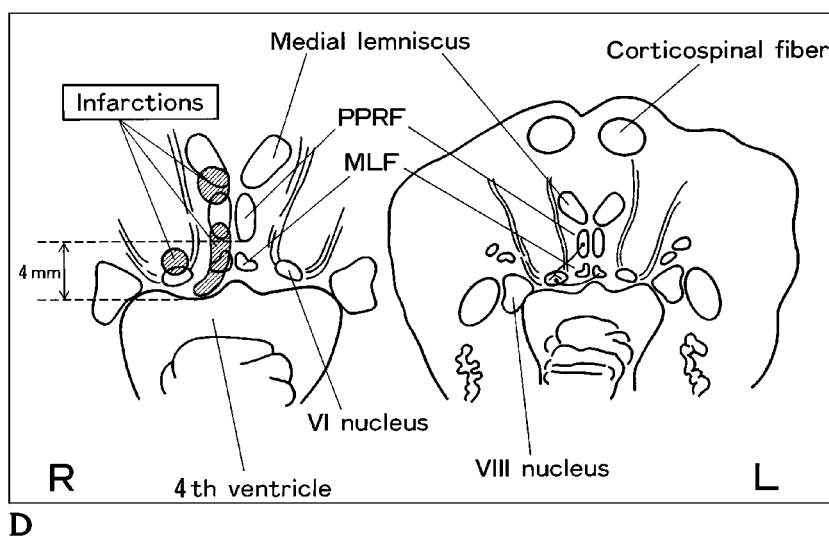
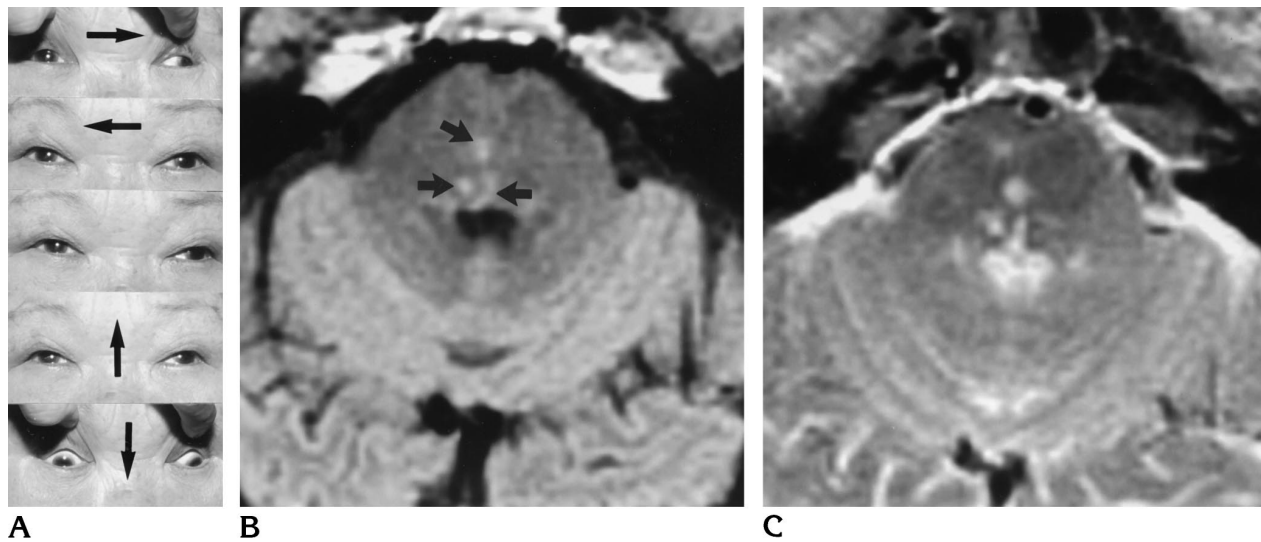


Fig 1. A, One-and-a-half syndrome in a 75-year-old woman with a pontine infarct. The *arrows* indicate the direction of attempted gaze. All ocular motor signs cleared within 1 month of onset.

B and C, MR images 3 days after the onset of the stroke showed three small, high-intensity lesions (*arrows*) in the dorsal portion of the right pontine tegmentum, contacting the floor of the fourth ventricle on proton density-weighted (2000/30/1 [repetition time/echo time/excitations]) (A) and T2-weighted (2000/100/1) (B) images.

D, Diagram of an axial section through the lower pons. The lesions (*shaded areas*) are on the right side (R), and the normal anatomy of the region and relevant structures are on the left side (L). Neurons project from the paramedian pontine reticular formation (PPRF) to the ipsilateral sixth-nerve nu-

cleus. Neurons in the sixth-nerve nucleus are both motor neurons, which represent the sixth nerve, and internuclear neurons, which ascend in the contralateral medial longitudinal fasciculus (MLF) (L). The lesions in this patient involved the MLF, PPRF, sixth-nerve nucleus, and medial lemniscus on the right side.

culus, PPRF, sixth nucleus, and medial lemniscus on the right side (Fig 1D).

Five days later, the horizontal gaze palsy toward the right side disappeared, and the dysesthesia and paresthesia of the corner of the mouth gradually reduced. The faint numbness of the left fingers and palm were still present. The residual right medial longitudinal fasciculus syndrome disappeared within 1 month of onset.

Discussion

The details of the MR and neuroanatomic findings of the one-and-a-half syndrome have been reported (6–8). The anatomic region in the lower pons, which includes the medial longitudinal fasciculus, PPRF, and sixth-nerve nucleus, is 10 mm in diameter.

Neurons project from the PPRF to the ipsilateral sixth-nerve nucleus. Neurons in this

nucleus are both motor, which represent the sixth nerve, and internuclear, which ascend in the contralateral medial longitudinal fasciculus (Fig 1D). Therefore, disease of either the sixth-nerve nucleus or the PPRF can cause a horizontal gaze palsy. Damage to the ipsilateral medial longitudinal fasciculus produces an internuclear ophthalmoplegia. Lesions that damage the medial longitudinal fasciculus on one side and the ipsilateral sixth-nerve nucleus (or PPRF) produce the one-and-a-half syndrome (1). In our patient, MR 3 days after the onset of the stroke revealed three small lesions in the dorsal portion of the right pontine tegmentum contacting the floor of the fourth ventricle (Fig 1B and C). These lesions involved the medial longitudinal fasciculus, PPRF, and sixth-nerve nucleus on the right side (Fig 1D).

Wall and Wray (5) have reviewed the reported cases of one-and-a-half syndrome and 20 cases of their own. They found that the most common causes of the syndrome were brain stem infarction (occurring in a variety of settings: hypertension, diabetes mellitus, atherosclerosis, cardiac disease, connective tissue disease, and polycythemia vera) and multiple sclerosis. These two entities have produced approximately two thirds of the reported cases. Other causes have included pontine glioma, arteriovenous malformation, pontine hemorrhage, basilar artery aneurysm, cerebellar astrocytoma, metastatic melanoma, and fourth ventricular ependymoma (5).

This patient had an associated cheirooral syndrome, which is typically caused by lesions located in the thalamus or cortex. Cheirooral syndrome caused by a pontine lesion has been recently reported (9–11). The etiology of the brain stem cheirooral syndrome is thought to involve the medial lemniscus, which extends horizontally on both sides of the paramedian pontine tegmentum (11). The ascending fibers from the lower limbs are

located on the outside; those from the upper limbs are located on the inside. The visceral pathway of the trigeminal nerve, which transmits the epicritic sensation of the face, ascends close to the rear inside of these fibers (11). In our patient, the lesions involved the inside of the right medial lemniscus in the lower pons (Fig 1D). In conclusion, a pontine infarct can produce both one-and-a-half syndrome and cheirooral syndrome.

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