Monocular elevation paresis and contralateral downgaze paresis from unilateral mesodiencephalic infarction

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Abstract
A 26 year old woman presented with monocular elevation paresis of the right eye, contralateral paresis of downward gaze, and subtle bilateral ptosis. Magnetic resonance imaging disclosed a unilateral embolic infarction restricted to the mesodiencephalic junction involving the left paramedian thalamus. Preserved vertical oculocephalic movements and intact Bell's phenomenon suggested a supranuclear lesion. This rare "crossed vertical gaze paresis" results from a lesion near the oculomotor nucleus affecting ipsilateral downward gaze and contralateral upward gaze fibres, originating in the rostral interstitital nucleus of the medial longitudinal fasciculus (riMLF).

Keywords: monocular elevation paresis; supranuclear lesion; mesencephalon

Jampel and Fells1 were the first to describe an acquired monocular elevation paresis, which was attributed clinically to a contralateral lesion in the pretectum. The hypothesis, that supranuclear fibres to the subnucleus of the oculomotor complex—which supplies the opposite superior rectus—are affected, has subsequently been confirmed by radiology and histopathology in a patient with similar symptoms.2 Its supranuclear nature is shown clinically by the persistence of vertical oculocephalic movements and the preserved Bell’s phenomenon. Our patient had an unusual combination of monocular elevation paresis and contralateral monocular paresis of downward gaze. Both radiological and clinical findings suggested a supranuclear lesion. To our knowledge, this type of vertical oculomotor dysfunction has not previously been reported.

Case report
A 26 year old woman noted a sudden onset of vertical diplopia, dizziness, and weakness of the right angle of the mouth. Ten months earlier an atrial septal defect had been diagnosed. One week before admission, the patient underwent cardiac catheterisation, which showed a bidirectional shunting of blood. There was no history of cerebral infarctions or other neurological diseases. Apart from a right sided mild supranuclear facial palsy, the neurological examination was normal. With the exception of a slight increase of LDH (248 U/l) and cholesterol (279 U/l) all laboratory studies gave normal results. Deep vein thrombosis was excluded by sonography. Duplex sonography of the carotid and vertebral arteries and carotid-vertebral Doppler ultrasonography were normal.

NEURO-OPHTHALMOLOGICAL FINDINGS
On admission mild bilateral ptosis was present, more pronounced on the left side. Both pupils were 4 mm in diameter and reacted normally to direct light and to near stimulus. Fundoscopy and visual acuity were normal and there was no history of strabism. In the primary position no deviation or cyclotorsion...
of the globe and fundus could be seen. On downward gaze and upward gaze, the patient mentioned vertical diplopia. Ductions showed a deficit of the left eye in downward gaze—more pronounced in left downward gaze—and of the right eye in upward gaze—equally severe with the eye abducted or adducted. This is consistent with underaction of the left inferior oblique and right superior oblique muscle. The right eye moved above the horizontal plane from 25° to 30°; the maximal amplitude of movement under the midline in the left eye was 20° in downgaze, and 10° in left downward gaze (fig 1).

Horizontal oculomotor ductions were of normal amplitude and convergence was present for both eyes. Oculocephalic movements were symmetrically preserved in both vertical directions and Bell’s phenomenon was normal.

Brain CT showed a small left paramedian thalamic hypodense lesion compatible with an infarction, involving the upper part of the left mesencephalon.

Magnetic resonance imaging of the brainstem disclosed an abnormal, high intensity signal on T2 weighted images 11 mm in diameter in the upper part of the left mesencephalon extending to the left ventromedial thalamic area. Figure 2A shows the sagittal T1 weighted SE image.

The patient received heparin intravenously for nine days. Diplopia, supranuclear facial palsy, and ptosis were greatly improved after two days, whereas dizziness persisted for one week. Brain CT showed a hypodense zone in the infarction area two weeks after onset. When the patient was last seen neurologically three weeks after admission, the elevation paresis on the right eye had almost disappeared. Downward gaze on the left eye was much improved as well, and the patient was able to depress the left eye about 30° to 35° under the midline. As the double vision was present only in left downward gaze, it was tolerated well by the patient. Follow up MRI one month after onset showed a sharply demarcated infarction zone of fluid like density (fig 2B). Two months after onset, the patient underwent surgery for the atrial septal defect.

**Discussion**

Monocular elevation paresis is regarded mainly as the so-called "double elevator
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Palpary,6 describing the inability to elevate one eye in all horizontal positions, equally severe in abduction or adduction. The acquired form of monocular elevation palsy was first described by Jampel and Fell. Because the superior rectus muscle is innervated by the contralateral superior rectus subnucleus and the inferior oblique is innervated by the ipsilateral inferior oblique subnucleus, they postulated that monocular elevation paresis of central origin is most likely due to a lesion of the supranuclear pathways for upward gaze in the contralateral pretectum. Subsequently only a few clinical cases have been reported, and the lesions were too large to localise precisely the impaired tracts.2 7

The critical areas involved in vertical gaze are the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF), the interstitial nucleus of Cajal, and the posterior commissure.8 Recent neurophysiological data (fig 3) showed that premotor neural medium lead burst neurons with downward on directions (DMLBs), similar to premotor neural medium lead burst neurons with upward on directions (UMLBs), are confined to the riMLF nucleus and that both types are intermixed. It is emphasised that the UMLBs of the riMLF ramify extensively within a restricted region of the oculomotor nucleus, corresponding to the territories of motor neurons supplying the inferior oblique and superior rectus muscles of both eyes. The projection of riMLF UMLBs to upward motor neuron pools was found to be bilateral. The RiMLF DMLBs, as well, ramify extensively within the dorsal two fifths of the rostral pole of the oculomotor nucleus and in the trochlear nucleus. By contrast with UMLBs, DMLBs have mainly ipsilateral projections to the oculomotor nucleus.14 15

If a lesion thus reaches the fibres after the decussation and before reaching the oculomotor nucleus, the elevation paralysis affects the contralateral eye. If the lesion affects the upgaze efferent fibres of the riMLF just after they leave the nucleus and before they decussate, an ipsilateral elevation palsy will result.10 In our patient the limitation of elevation was equally severe with the eye abducted or adducted, suggesting a palsy of the superior rectus muscle, according to the predominance of action of the superior rectus in all positions of gaze. The lesion of the meso diencephalic junction thus may have destroyed premotor fibres projecting on to the ipsilateral superior rectus nucleus (fig 3).

Preserved Bell’s phenomenon and oculocerephalic movements as well as the radiological findings are strong arguments for a supranuclear lesion. The mild bilateral ptosis, being more severe on the left side, accounts for involvement of the central causal nucleus.

To our knowledge, monocular palsy of downward gaze—as shown in our patient—has been reported only twice.11 12 In the first case11 destruction of fibres ascending from the left vestibular nuclei to the nucleus that innervates the left inferior rectus was suggested. In the second report,12 as could be the case in our patient, it was assumed that descending fibres from the riMLF to the ipsilateral subnucleus of the inferior rectus were destroyed. In our patient the downgaze paresis on the left eye was more pronounced when the eye was abducted, which accounts for an isolated palsy of the inferior rectus.

Monocular elevation paresis, contralateral downward gaze palsy, and mild bilateral ptosis can theoretically be caused by a lesion in one oculomotor nucleus. Clinically, however, this would at least coincide with loss of Bell’s phenomenon and upward oculocephalic responses, which was not the case in our patient. Furthermore, it is difficult to make a strong argument for a monocular lesion in the absence of any pupillary mydriasis, accommodative dysfunction, medial rectus involvement, or more impressive ptosis.

These findings support the possibility of a monocular elevation palsy and contralateral downward gaze paresis due to a unilateral mesodiencephalic lesion, as presented in our patient. This rare case of disconjugate oculomotor disorder provides further evidence for the supranuclear organisation of vertical gaze.
