Conjugate upward gaze paralysis with unilateral ptosis caused by a unilateral midbrain infarction

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A 73-years old woman with atrial fibrillation presented with a sudden right hemiparesis, with diplopia and left ptosis, and was admitted at an Emergency Unit. The neurological examination showed fluctuations on consciousness level, predominant crural right hemiparesis, right central facial paralysis without sensitive abnormalities. The first ophthalmological evaluation showed normal pupillary reflexes, total left ptosis, paresis of adduction of the left eye, with conjugated horizontal palsy for right gaze and conjugated vertical palsy for upward and downward gaze on saccadic and smooth pursuit eye movements. The convergence showed paresis of left eye, with reactive pupils, and oculocephalic test was normal. A head computed tomography (CT) had no acute ischemic signs, and after four days, she was discharged. The brain magnetic
resonance (MR) performed 15 days after the ictus showed a clearly defined left paramedian tegmental mesencephalic infarct (Figure 1). Two months after the stroke, the patient had a remarkable improvement of ocular motility, presenting paresis of levator palpebrae, medial and inferior rectus muscles of the left eye, with conjugate vertical upward gaze palsy in saccadic and smooth pursuit (Figure 2).

The control of the vertical gaze within the brainstem is mediated by three main nuclei: the nucleus of rostral interstitial medial longitudinal fasciculus (riMLF), the interstitial nucleus of Cajal (INC) and nucleus of the posterior commissure (NPC) [1]. In primates, there is a coordinated action among the three nuclei for vertical gaze generation, but the INC and NPC have a main role in upward eye control [1]. The riMLF/INC/NPC system projects its axons, through the medial longitudinal fasciculus (MLF), to the oculomotor complex (OC) by distinct pathways: in upward gaze, these fibers innervate both ipsilateral and contralateral elevator muscles subnuclei (rectus superior and oblique inferior) of OC simultaneously; in downward gaze, the projections to rectus inferior and oblique superior run predominantly ipsilateral [1, 2]. There are important connections between these MRF nuclei, and due its proximity, small lesions of MRF can affect all of them [2, 3]. Ptosis is caused by a lesion in central caudal nucleus (CCN) neurons or its fibers. The CCN innervates both levator palpebrae muscles [1]. Classically, midbrain lesions affecting the CCN lead to bilateral ptosis [4], but partial lesions of the oculomotor fascicle, situated in the paramedian ventral midbrain, can cause unilateral ptosis [5].

Our patient had an unusual association of neurological abnormalities. This combination of eye movement disorders and right hemiparesis could be explained by a left paramedian midbrain lesion, at the level of: (1) the riMLF/INC/NPC system fibers projecting to the OC (mainly the INC and NPC axons), causing the vertical gaze palsy, (2) left oculomotor fascicle, associated with the total left ptosis with medial and inferior rectus palsies, sparing the pupil, and (3) left cerebral peduncle, explaining right hemiparesis (Figure 3). There are others descriptions of unilateral
midbrain lesions causing supranuclear vertical gaze paralysis [3, 6, 7, 8], but no case had unilateral ptosis. This case shows the complexity of supranuclear eye movement control and the intrinsic relationship among the midbrain reticular formation nuclei.

FOOTNOTES

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REFERENCES


LEGENDS

Figure 1 – MR images from the case. A: Axial T2 FLAIR. B: Axial diffusion weighted MR image. C: ADC map, showing a small acute infarction (arrow) in the left paramedian tegmental midbrain.

Figure 2 – Positions of gaze. A: Forward gaze in primary position, with elevation of eyelids. B: Forward gaze in primary position (showing left ptosis). C: Upward gaze (with bilateral conjugated upward gaze palsy). D: Downward gaze (showing downward paresis of left eye). E: Right gaze (with adduction paresis of left eye). F: Left gaze.

Figure 3 – Anatomic scheme of brainstem nuclei associated with control of eye movement, in sagittal and axial view. A: Structures of vertical gaze control, in sagittal view, with two levels (B and C) delimited through midbrain. B and C: Axial sections from the levels indicated in A. The shaded area indicates the presumed site of lesion. 3N, oculomotor nucleus; 3f, oculomotor fascicle; 3n, oculomotor nerve; 4N, trochlear nucleus; 6N, abducens nucleus; 6f, abducens fascicle; CCN, central caudal nucleus; CP, cerebral peduncle; CTG, central tegmental tract; INC, interstitial nucleus of Cajal; MLF, medial longitudinal fascicle; MmB, mammilary body; OC, olivary complex; PC, posterior commissure; rMLF, rostral interstitial nucleus of the medial longitudinal fascicle; SC, superior colliculus (figure modified from [1]).
MR images from the case. A: Axial T2 FLAIR. B: Axial diffusion weighted MR image. C: ADC map, showing a small acute infarction (arrow) in the left paramedian tegmental midbrain.

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MULTIPLE CHOICE QUESTION - IMAGE QUIZ

A 73-years old woman presented suddenly a right hemiparesis, with left ptosis, left medial and inferior rectus muscles palsy, and a conjugated upward vertical gaze palsy (Figure 2). MR image showed a left paramedian tegmental mesencephalic infarct (Figure 1). Which is the most common affected artery on this topography?

Options:

a) Superior cerebellar artery
b) Posterior thalamo-subthalamic paramedian artery
c) Posterior communicating artery
d) Anterior choroidal artery
e) Distal branches of middle cerebral artery

Correct answer:

b) Posterior thalamo-subthalamic paramedian artery

Explanation: Midbrain infarcts occur in 1% of all ischemic strokes, and isolated midbrain ischemic strokes are found in 0.7% of all posterior circulation infarcts [1]. The most common affected vessel is the posterior thalamo-subthalamic paramedian artery, a branch of the basilar artery on its top, and the unilateral lesion is more frequent in midbrain strokes [2], which also can be described as medial mesencephalic branches of top of basilar [3]. Reporting 21 cases of isolated
midbrain infarct, Ogawa et al. noted that the paramedian region was the most affected area of midbrain, causing a multitude of clinical signs, as hemiparesis, ataxia, eye movement disorders, including ptosis and pupil defects [3].

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