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ONE AND HALF SYNDROME – A CASE REPORT

Medical Science

KEYWORDS : Medial longitudinal fasciculus, pontine infarct ,Internuclear ophthalmoplegia .

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ABSTRACT

We report a case of 29 year old female who presented with features of sudden onset ptosis of right eye with gait instability and diplopia. Clinical examination revealed ocular position of the right eye on forward gaze fixed at the midline, while the left eye was abducted. On rightward gaze, neither eye could pass the midline, and on leftward gaze the left eye was abducted with monocular nystagmus suggestive of internuclear ophthalmoplegia.

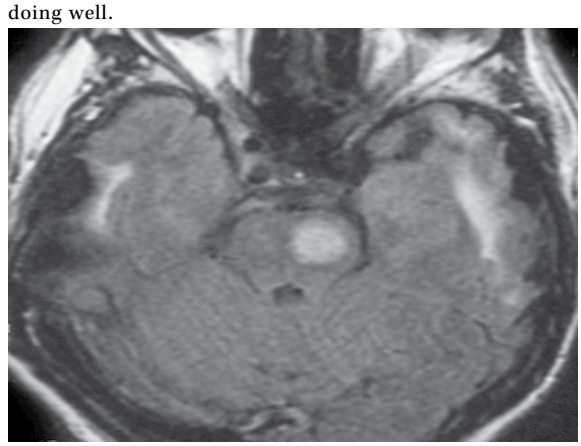
Introduction

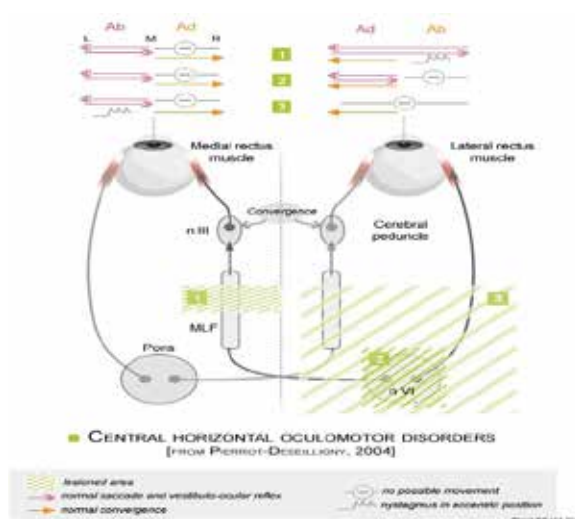
The one-and-a-half syndrome is a clinical disorder of extraocular movements characterized by a conjugate horizontal palsy in one direction plus an internuclear opthalmoplegia in the other. It is due to a unilateral lesion of the dorsal pontine tegmentum, involving the ipsilateral paramedian reticular formation, internuclear fibers of the ipsilateral medial longitudinal fasciculus and, usually, the abducens nucleus. The main causes of this rare syndrome are stroke and multiple sclerosis. Other causes include tumors, AV malformations, basilar artery aneurysms and rarely, vasculitis, brainstem tuberculoma and neurocysticercosis.

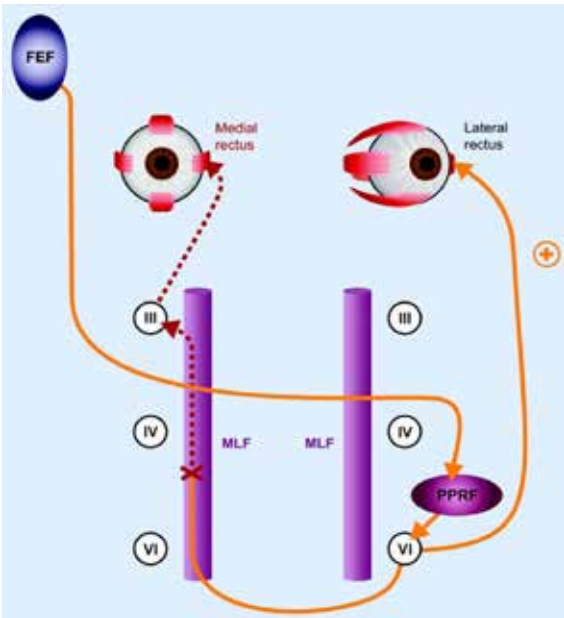
CASE REPORT-

A 29-year-old female patient was admitted to our institution with sudden onset of drooping of right eyelid, vertiginous feeling, gait instability and diplopia. She denied any history of weakness of any part of the body, paresthesias or numbness of limbs or face, urinary incontinence, deafness, tinnitus or any slurring of speech. She volunteered that she experienced sudden onset drooping of her right eyelid with diplopia. She has no past history of being hypertensive or diabetic. On admission, she had Glasgow coma score of 15/15 and her blood pressure was 140/90 mmHg. Her pulse was regular and equal on both sides with all peripheral pulses present. The ocular position of the right eye on forward gaze was fixed at the midline, while the left eye was abducted. For horizontal ocular movements, only the left eye could abduct with monocular horizontal nystagmus. Also, there was conjugate gaze palsy to right, indicating horizontal right one-and a-half syndrome i.e. on rightward gaze, neither eye could pass the midline, and on leftward gaze, horizontal jerky nystagmus in the abducting left eye was noticed. Vertical and rotatory nystagmus was absent.

There was no diplopia in primary gaze but she experienced horizontal crossed diplopia in leftward gaze. Vertical eye movements were preserved. Visual acuity was 6/6 (Snellen) and N/6 in both eyes. Pupillary reactions were normal. The fundi were normal with no evidence of papilledema. No other neurologic deficit was seen. Rest of the systemic examination was also inconclusive. Her main symptom at this time was oscillopsia due to the abducting nystagmus of her on-paretic right eye, which was only relieved with occlusion of the eye with a patch. A clinical diagnosis of one-and-a-half syndrome secondary to a cerebrovascular event was made. And MRI of the brain showed small focal area of altered intensity on posterior aspect of pons suggestive of acute infarct. Patient was managed conservatively with anticoagulants with a regular check on her vitals in view of the strategic location of infarct. Over the days, the patient became symptomatically better although oscillopsia due to abducting nystagmus of her non-paretic left eye still persisted. She is on regular neurology follow up and is doing well.







DISCUSSION

In 1967,Miller Fisher(9) described a syndrome,he termed "one-and-a-half-syndrome" (OAHS) consisting of a "conjugate lateral gaze palsy in one direction, plus one half of a gaze palsy in the other". Many aetiologies of OAHS have been described(1,2) mostly vascular, but also demyelinating and neoplasic disorders. The syndrome usually results from a single, unilateral and relatively small lesion at the dorsal tegmentum of the lower pons. Occasionally an ocular paralysis occurs, one eye looking ahead without any horizontal movement and the other staying abducted. The latter eye does not move beyond the midline when horizontal movements are tested. Vertical eye movements are relatively preserved. In OAHS the abducens nucleus (AN) and/or the paramedian pontine reticular formation (PPRF) located rostrally and ventrally to that nucleus are damaged, producing an ipsilateral conjugate gaze palsy. Damage to the ipsilateral medial longitudinal fasciculus (MLF), which lies at the same level or just above the AN, accounts for the unilateral internuclear ophthalmoplegia (INO).In addition, during the acute phase, exotropia of the contralateral eye, no deviation, or less commonly, esotropia of the eye ipsilateral to the lesion secondary to the paresis of the sixth nerve, may be seen in primary position of gaze. The present patient had a deficit of horizontal gaze to the right side combined with a paresis of

adduction to the opposite direction. In this syndrome, the normal vertical eye movements and convergence suggest that motor fibers and nuclei of the third cranial nerves were preserved. The degree and duration of the horizontal gaze impairment caused by unilateral lesions in the brain above the level of the oculomotor nucleus may vary according to the size and location of the lesion.The abducens nucleus (AN): extensive experimental data from cats and monkeys(3 4) suggest that the AN contains two types of cells, i.e., motoneurons which innervate the ipsilateral external rectus muscle (magnocellular motor neurons) and excitatory internuclear neurons that decussate at the level of the nucleus, ascend in the contralateral MLF and terminate at the medial rectus subdivision of the oculomotor nuclear complex. The paramedian pontine reticular formation (PPRF): numerous experimental studies in monkeys have produced data supporting the importance of the PPRF for integration of horizontalconjugate gaze (4,6 8 10). Smaller lesions (1 to 2 mm) in the tegmentum of the pons, specifically in the PPRF, induce paralysis of ipsilateral conjugate gaze for as long as one year.

Outside the pons the amount of tissue destruction necessary to affect horizontal gaze is much greater than in PPRF lesions(10). The PPRF contains two main types of neurons associated with eye movements, the phasic and the tonic cells. The former (excitatory burst neurons) are active during saccades, namely all kinds of rapid eye movements (voluntary saccades and quick phases of nystagmus).

Intemuclear ophthalmoplegia (INO): the INO is characterized by paresis or paralysis of the ipsilateral eye adduction - on attempted horizontal gaze to the contralateral side - and horizontal jerk nystagmus in the contralateral abducting eye. Typically convergence is preserved if the lesion does not extend to the mesencephalon. Bilateral lesions cause bilateral defects in adduction and nystagmus in the abducting eye. In addition there are nystagmus on upward gaze, as well as bilateral partial ptosis. Lesions of the MLF do not cause paralysis of conjugate horizontal or vertical gaze(7). Upgaze palsy, downgaze palsy, complete vertical gaze palsy and vertical saccade for both upward and downward gaze have been known to be related to bilateral lesions involving the rostral interstitial nucleus of medial longitudinal fasciculus (RiMLF) cell group and the posterior commissure(5).Paralytic pontine exotropia (PPE): the presence of exotropia in OAHS was first observed by Fisher(9) In the acute phase, the exotropic eye shows abduction nystagmus during attempts to move it further laterally, and there is extreme slowness of adduction when the eye moved to the midline.Again, normal midbrain ocular motor functions are shown by intact vertical gaze, convergence, and pupillary constrictor reflex activity. In conclusion, the semiological diagnosis of the OAHS is comparatively simple and allows great anatomical precision. Peripheral facial palsy without gustatory deficit is a common associated sign and eventually the most exuberant symptom.

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