Case Report

One and a half syndrome

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Abstract

The one and a half syndrome is a rare ophthalmoparetic syndrome characterized by "*a conjugate horizontal gaze palsy in one direction and an inter-nuclear ophthalmoplegia in the other*". The most common manifestation of this unusual syndrome is limitation of horizontal eye movement to abduction (moving away from the midline) of one eye with no horizontal movement of the other eye. Nystagmus is also present when the eye on the opposite side of the lesion is abducted. Convergence is classically spared as Cranial Nerve III (oculomotor nerve) and its nucleus is spared bilaterally.
Key Words: Nystagmus, oculomotor nerve.

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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 internuclearophthalmoplegia in the other. It is due to a unilateral lesion of the dorsal pontinetegmentum, involving the ipsilateral Paramedian Pontine Reticular Formation (PPRF), internuclearfibers of the ipsilateral medial longitudinal fasciculus (MLF) (responsible for ipsilateral failed adduction) and, usually, the abducens nucleus(responsible for horizontal gaze palsy). 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. When this lesion also involves the fascicle of ipsilateral facial nerve (Cranial Nerve VII) in the region of the facial colliculus as it wraps around the CN VI nucleus, it produces a lower motor neuron pattern of ipsilateral facial weakness, it is called Eight-And-A-Half Syndrome. (One-and-a-half syndrome plus 7thCranial nerve palsy)

CASE REPORT

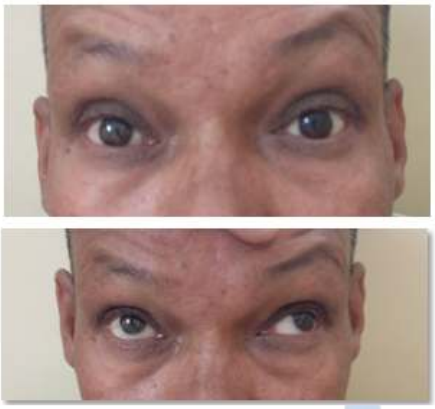
A 52 years Male patient was referred from Medicine department for evaluation of diplopia and left eye ptosis. However he denied any history of weakness of any part of the body, paresthesias or numbness of limbs or face, deafness, tinnitus, slurring of speech or urinary incontinence. He volunteered that he suddenly noticed drooping of left eyelid with diplopia. He is a well controlled case of Diabetes Mellitus for 5 years on oral hypoglycaemic medications.

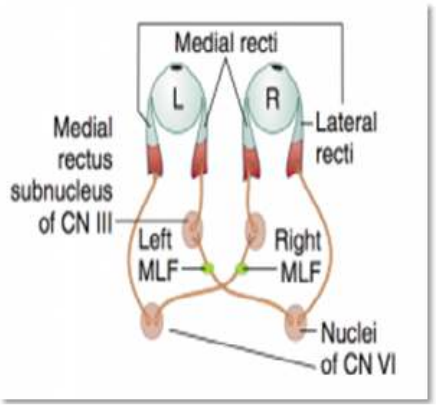
Ophthalmological Examination

The patient has unaided visual acuity of 6/24 and 6/18 respectively in right and left eye. Anterior segment examination revealed Moderate degree of ptosis and Horizontal Jerky Nystagmus on abduction in left eye. Corneal sensations were normal in both eyes. Pupillary reactions were normal with Immature cataract in both eyes. The ocular position of the right eye on forward gaze was fixed at the midline, while the left eye was slightly abducted. For horizontal ocular movements, only the left eye could abduct with horizontal nystagmus, right eye has restriction of movements in horizontal gaze. There was no diplopia in primary gaze but she experienced horizontal crossed diplopia in leftward gaze. The fundi were normal with no evidence of papilledema.

Neurological Examination

There is nothing suggestive of involvement of Cranial Nerve VII (Facial Nerve). No other neurologic deficit was seen. A clinical diagnosis of one-and-a-half syndrome secondary to a cerebrovascular event was confirmed by MRI of the brain showing small focal area of altered intensity on posterior aspect of pons suggestive of acute infarct. The patient was managed conservatively on Anticoagulants with regular follow-ups with Medicine and Neurology.





DISCUSSION

In 1967, Miller Fisher 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 etiologies of OAHS have been described mostly vascular, but also demyelinating and neoplastic disorders. The syndrome usually results from a single, unilateral and relatively small lesion at the dorsal tegmentum of the lower pons. 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 abducensnucleus (AN) and/or the paramedianpontine reticular formation (PPRF) locatedrostrally and ventrally to that nucleus are damaged, producingan ipsilateral conjugate horizontal 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 internuclearophthalmoplegia (INO). 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) contains two types of cells, i.e., motorneurons which innervate the ipsilateral external(Lateral) 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) is important for integration of horizontal conjugate 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. 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 all kinds of rapid eye movements (voluntary saccades and quick phases of nystagmus). Intemuclear Ophthalmoplegia (INO) is characterized by paresis or paralysis of the ipsilateral eye adduction - on attempted horizontal gaze to the contralateral side and horizontal jerky 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. Normal midbrain ocular motor functions are shown by intact vertical gaze, convergence, and pupillary constrictor reflex activity.

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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