CASE REPORT

WALL-EYED MONOCULAR INTER-NUCLEAR OPHTHALMOPLEGIA (WEMINO) SYNDROME, A RARE DECISIVE MANIFESTATION OF AN ISOLATED UNILATERAL PONTINE STROKE

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Isolated unilateral pontine infarction is an uncommon occurrence. It may bring a complex neuro-ophthalmic manifestation to the clinicians, making the on-spot-diagnosis a hard challenge. Wall-eyed monocular inter-nuclear ophthalmoplegia (WEMINO) syndrome is a rare variant of inter-nuclear ophthalmoplegia which also includes ipsilesional exotropia. The literature seems deficient in documenting WEMINO syndrome the primary presentation of isolated unilateral pontine stroke. The location of the causative lesion of WEMINO syndrome is a hot topic. Here, we discuss a case of WEMINO syndrome, a rare presentation of limited unilateral pontine stroke and its responsible lesion. To the best of our knowledge this is the first reported case from Pakistan. A short literature review has also been presented on the anatomy, pathophysiology and various manifestations of isolated unilateral pontine lesion in the region of medial longitudinal fascicules (MLF). Hence, this article enhances the understanding of clinicians regarding the responsible lesions limited to pons and its various manifestations, in order to enable clinicians to pick them in the early opportunity.

Keywords: Ophthalmoplegia; WEMINO; INO; Pontine stroke; MLF lesion

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INTRODUCTION

Isolated pontine stroke represents a minority of all ischemic strokes. It often presents in a complex and challenging way to the physicians, because of the heterogeneity of presenting phenotypes.[1,2] There is a deficiency of reported cases in the literature regarding an isolated unilateral pontine lesions and their various neuro-ophthalmic presentations.[3] Similarly, a unilateral Inter-nuclear ophthalmoplegia with homolateral exotropia (WEMINO syndrome) secondary to an isolated pontine stroke is a rare occurrence and reported extreme rarely.[2] Only few cases have been reported, describing the WEMINO syndrome and its radiological findings.[3]

Wall eyed monocular inter-nuclear ophthalmoplegia (WEMINO) syndrome is a unilateral inter-nuclear ophthalmoplegia and ipsilateral exotropia associated with an ipsilateral medial longitudinal fasciculus (MLF) infarction.[2,4] Here, we have reported a rare case of WEMINO syndrome and its neuro-radiological evidence.

CASE PRESENTATION

A 36-year-old woman with a history of hypertension for the last 10 years, presented to the emergency department with the complaints of sudden onset of headache, dizziness, blurred vision and diplopia. It was six hours after the onset of symptoms and she was mainly concerned regarding her blurred vision and the diplopia, which was binocular and both in the horizontal and vertical gazes. She had an episode of vomiting that day, however denied fever, photophobia, vertigo, hearing loss, dysarthria, dysphagia, loss of consciousness or any kind of head trauma. Her both eyes were previously normal.

Despite a significant family history of essential hypertension in the early age, the family history was unremarkable for cerebrovascular and cardiovascular accidents. Her medications included losartan 50mg and hydrochlorothiazide 12.5 mg. The patient was admitted for further assessment and hypertension control.

On physical examination, her blood pressure was 260/130 mmHg in both arms with a pulse rate of 113 per min, Temperature of 98F and Respiratory rate of 14. The patient was well oriented and she was responding normally to verbal commands. There were no focal neurological signs including speech, motor, sensory or cerebellar signs. Apart from ocular movements, the cranial nerves examination was unremarkable.

Ophthalmological examination revealed, unaided visual acuity of 6/36 in both eyes with normal reactive pupils. On neuro-ophthalmic assessment, the left eye in primary position was deviated outward and upward with no head tilt (40 dioptres exotropia and 14 dioptres hypertropia on prism cover test). On her right lateral gaze, the left eye failed to adduct beyond the midline, and the right eye had abductive nystagmus with the fast component toward the midline (video: https://vimeo.com/519405793). On her left lateral gaze,

the left eye had some limitations in abduction. An anticlockwise (defined with respect to the patient) torsional nystagmus was appreciated on horizontal saccades. On vertical gaze, misalignment could be appreciated due to left hypertropia. The left eye had limitation in convergence also. All these features conclude Left Inter-nuclear ophthalmoplegia with ipsilateral exotropia and hypertropia (WEMINO syndrome). The fundus was normal on ophthalmoscopy.

The initial CAT scan of brain in the emergency department revealed no pathology. An MRI scan (1.5 Tesla GE Machine) of the brain without IV contrast was performed on the 3rd day of admission. T1WI, T2WI, FLAIR, DWI/ADC 5mm images in sagittal, coronal and axial slices were acquired. A tiny area (3.5mm- isointense on T1WI and hyper-intense on T2WI) of restricted diffusion suggestive of acute infarct, in the left dorsal pons in the region of MLF anterior to the 4th ventricle was noted. (Figure-3).

She was also investigated for secondary cause of hypertension, which was not found and therefore, essential hypertension was concluded. Subsequently, she was treated as an in-patient in internal medicine unit. In the course of seven days after admission, her hypertension was controlled on quadruple therapy (losartan, amlodipine, hydrochlorothiazide and doxazosin) and the ocular movements showed little improvement. The diplopia in the left lateral gaze and the exotropia in the left eye begun to improve. The left eye was able to cross the midline, but failed to adduct fully.

On follow up after 8 weeks, the patient was re-examined in the internal medicine unit OPD in collaboration with ophthalmology unit. Her vitals were stable with blood pressure 130/80 mmHg and pulse 82 bpm. She admitted improvement in diplopia except on the right upward gaze. The assessment of eye movements revealed a significant improvement in the left exotropia, adduction in the left eye and convergence (Figure-2).

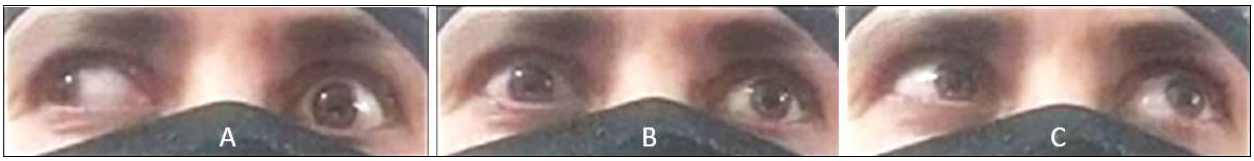

Figure-1: At the time of discharge from the hospital, (B) the left exotropia with hypertropia started to resolve in the primary gaze. (A) On right lateral gaze, the left eye fails to adduct fully. (C) On left lateral gaze, the left eye shows some limitation in abduction.

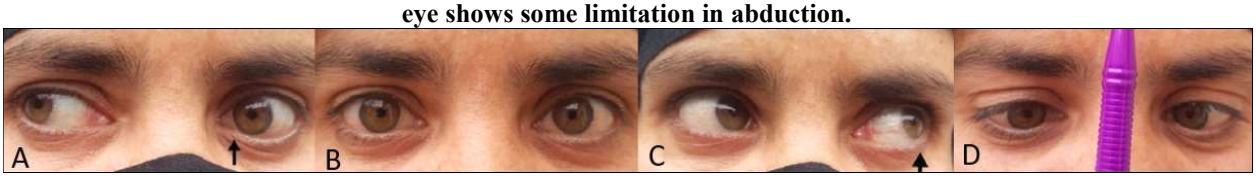

Figure-2*:* (B) In primary gaze the left exotropia has resolved. (A) In the right lateral gaze, the left eye adducts with minimal limitation as seen in the photograph. (C) In the left lateral gaze, the left eye hypertropia can be appreciated. (D) The convergence is adequate.

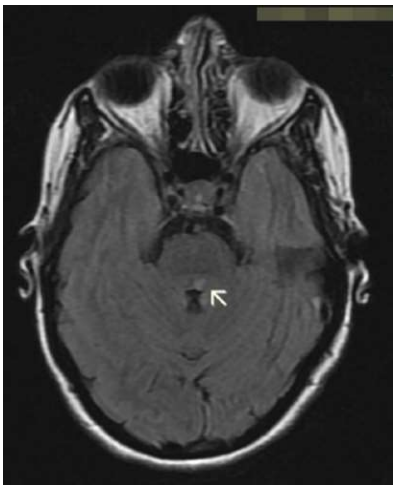

Figure-3: The area pointed by the small white arrow corresponds to a small area of restricted diffusion unilaterally at the left, upper dorsal region of pons.

DISCUSSION

Stroke accidents involving only the pontine region, happen rarely.[2] The presenting phenotype is often variable[5] and is therefore, often misdiagnosed. WEMINO syndrome and the radiological findings of it causative lesion have been reported only rarely.[3,4] Our patient presented with INO and ipsilateral exotropia and hypertropia. On MRI scan (T2 and Flair) a tiny hyper-intense lesion was found in the left MLF region of pons. The radiological finding of unilateral MLF region lesion associated with this rare presentation has been reported once by Ikeda *et al*.[3]

Pons, a complex integral part of brainstem, is structurally and functionally divided into ventral part and dorsal part (tegmentum). The later one lies anterior to the fourth ventricle containing ascending and descending projections, certain cranial nerves nuclei (V to VIII) and the extremely important MLF.[2]

Medial rectus and the lateral rectus muscles execute the horizontal ocular movements. They are innervated by the oculomotor nerve (III cranial nerve) and the abducens nerve (VI cranial nerve), respectively. The oculomotor and the abducens nuclei are interconnected by a tract in the brainstem named the MLF, which coordinates them generating a horizontal conjugate eye movement. Paramedian pontine reticular formation (PPRF) in the pons acts as the interconnecting structure which initiates the horizontal conjugate eye movement.[6] Any lesion of the MLF causes a discrete adduction failure of ipsilateral eye and an abduction nystagmus in the contralesional eye during horizontal gaze, which is termed as an INO.[5] Our patient was a young lady who had left INO with ipsilateral exotropia and ipsilateral hypertropia with the only risk factor for stroke as hypertension, made it challenging for us to conclude stroke at the early opportunity. The pontine disorders manifest in a wide range of clinical phenotypes depending on the involvement of different arterial territories.[2] Although, each arterial territory is exclusively supplied by certain artery[7], the clinical manifestations are not always absolutely discrete[8]. Ocular motility disorders is one of the many kinds of primary pontine stroke manifestations. WEMINO syndrome, so far reported in literature, has different additional ocular motility disorders as well. For example, a case of WEMINO was reported with the addition of upward nystagmus associated with the same pontine lesion (MLF region pontine stroke).[3] An exactly same case of WEMINO syndrome was reported by Jeon (2005) who had additional upbeat nystagmus, which was lacking in our patient. The combination of INO with contralesional exotropia is not uncommon.[9] which is defined as Non-paralytic pontine exotropia.[10] Johkura and Kudo (2015) reported that 56% of the patients with Unilateral INO had contralateral exotropia. But the reason of underreporting of contralateral exotropia with INO is possibly, because it tends to be transient and often overlooked.[9] On the other hand, an isolated unilateral pontine stroke can present in other ways as well: Zhang and Xio (2015)[11] reported a case of limited pontine stroke (right MLF region of dorsal pons) in which they reported only right INO with bilateral upbeat nystagmus in the primary position, but without exotropia.

In the WEMINO syndrome the INO could be explained by ipsilateral pontine stroke around the MLF[2,6], however, the exact description of the lesion responsible for the ipsilesional exotropia still remains unclear as the limited tegmental damage fails to explain that the exotropia could be attributed to oculomotor nerve paresis.[2,3] This illustrates that, a tiny unilateral pontine stroke (limited to MLF region) could present in three possible ways:

- Ipsilateral INO without exotropia
- Ipsilateral INO with contralateral exotropia
- Ipsilateral INO with ipsilateral exotropia (WEMINO syndrome)

A larger lesion at the MLF region may also involves the nearby structures including ipsilateral PPRF and abducens nucleus which further complicates the presentation, defined as one and a half syndrome (Lateral gaze palsy in ipsilateral direction and INO in the contralateral gaze).[12] The genu of facial nerve (VII cranial nerve) lies around the abducens nucleus in the dorsal pons. The lesion responsible for one-and-a-half-syndrome may extend to involve the genu of facial nerve (VII cranial nerve), resulting in eight-and-a-half syndrome, which is one-and-a-half-syndrome with ipsilesional lower facial nerve palsy (VII cranial nerve).[13]

CONCLUSION

A pontine stroke can present in a variety of ways and often implies a challenge for the clinicians including specialists. The findings in our case were typical of WEMINO syndrome which has not been fully addressed in the literature. The constellation of Unilateral INO with ipsilateral exotropia and hypertropia are suggestive of a tiny unilateral MLF infarction which is often missed on MRI. The clinicians must be aware of this clinical presentation and should be ready to address it timely.

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