

Original Research Article

A rare case of eight and a half syndrome due to pontine hemorrhage - A case report

Sandeep Reddy Nareddy¹, E.A. Ashok Kumar^{2*}

¹Senior Resident, ²Professor

Department of General Medicine, Malla Reddy Institute of Medical Sciences, Hyderabad, India

*Corresponding author email: ashokedla@gmail.com

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Abstract

Eight and a half syndrome includes both one and half syndrome and peripheral facial paralysis. One and a half syndrome, which was named by C Miller Fisher in 1967, includes ipsilateral horizontal gaze palsy (one) and ipsilateral Inter Nuclear Ophthalmoplegia (INO) (a half). This condition is caused by circumscribed lesions of the Pontine Tegmentum and if the lesions expand to the adjacent structures, various other associated clinical manifestations may appear based on the underlying involved anatomical structures. We report a case of 50 year old male who presented with Eight and a Half Syndrome associated with ipsilateral ataxia, due to pontine hemorrhage.

Key words

Eight and a Half Syndrome, Pontine Tegmentum, Pontine Hemorrhage.

Introduction

The pontine tegmentum contains nuclei of the cranial nerves, abducens, facial, and vestibule cochlear and their associated fibre tracts, the Pontine Paramedian Reticular Formation (PPRF) and the medial longitudinal fasciculus and the involvement of these structures could result in gaze palsy, INO, nystagmus, peripheral facial nerve palsy [1]. Freeman, et al., described a combination of horizontal gaze palsy along with INO due to lesion in PPRF and MLF in 1943 [2], later C Miller Fisher in 1967 coined the term one

and a half syndrome [3]. We herewith reported a case of Eight and a half syndrome, which includes one and half syndrome along with LMN facial palsy and left cerebellar dysfunction.

Case report

A 50 year old male presented with history of sudden onset of nausea, vomiting, giddiness and headache followed by horizontal diplopia which is more on looking to right side, patient also had left side facial weakness with difficulty in closing the eyelid and deviation of angle of the

mouth to the right and swaying to the left side. Patient had ischemic stroke in 2016 with left hemiparesis from which he recovered completely. His medical history was significant for history of ischemic stroke, hypertension. He was on irregular medications and is not taking antiplatelets and antihypertensives since the past three months.

General physical examination was unremarkable, Temp – Normal, Resp. Rate – 16/mt, PR 79/min regular in rhythm, Blood Pressure of 200/120 mmHg.

On CNS examination - Pupils were equal and reactive to light, Oculomotor abnormalities were observed in the form of left horizontal gaze palsy i.e., failure of left eye abduction and right eye adduction and there was no horizontal

movements in left eye with abducting nystagmus of right eye, the only lateral ocular movement that remained was abduction and nystagmus of right eye i.e., suggestive of left INO lesion (**Figure - 1**). The left eye abduction weakness was much more than the adduction weakness resulting in adducted eye in neutral position and possible involvement of abducens nucleus rather than Paramedian Pontine Reticular Formation as the mechanism of left horizontal gaze palsy in this case. Vertical eye movements were preserved. Left lower motor neuron facial palsy was observed. All other cranial nerves were normal. Motor and sensory system examination was normal. On cerebellar examination, body was swaying to left side while walking in a straight line, left sided finger – nose and heel-knee tests were normal. Other systems examination was normal.

Figure – 1: A) On left lateral gaze, B) On right lateral gaze, C) On upward gaze, D) On downward gaze, E) Left LMN facial palsy.

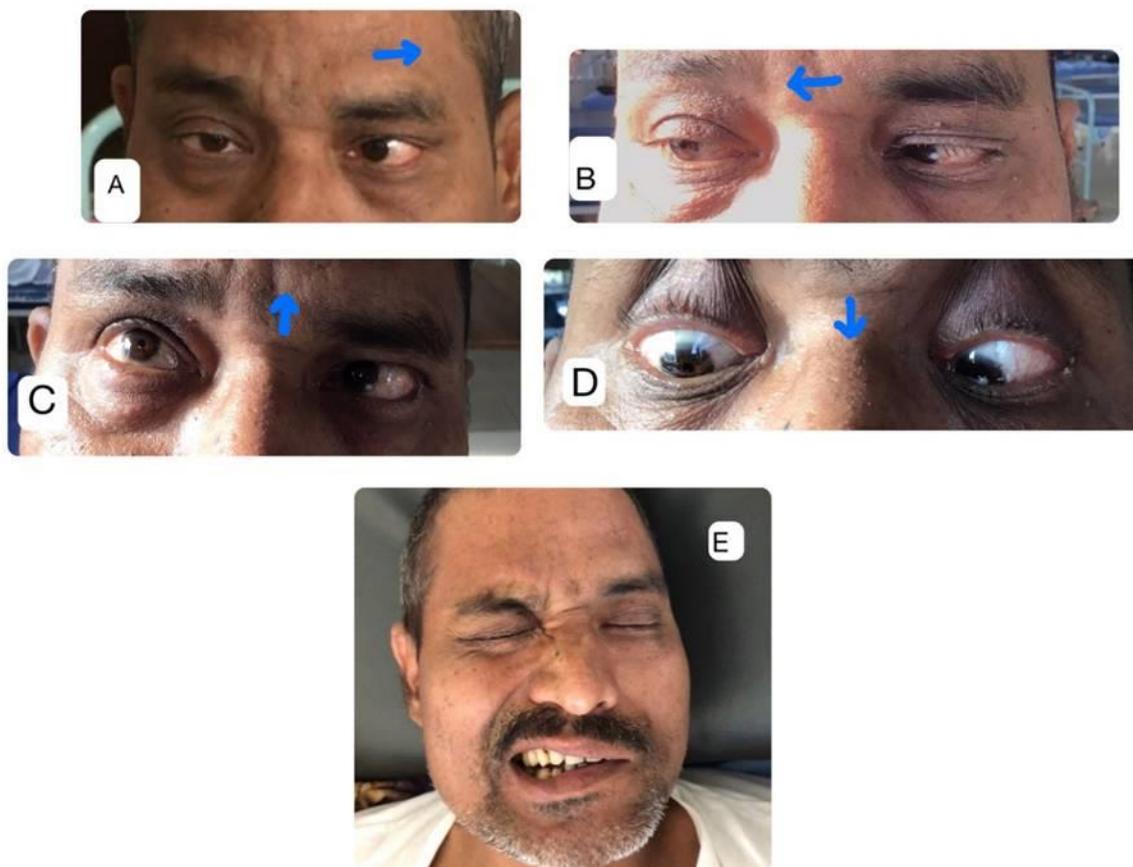
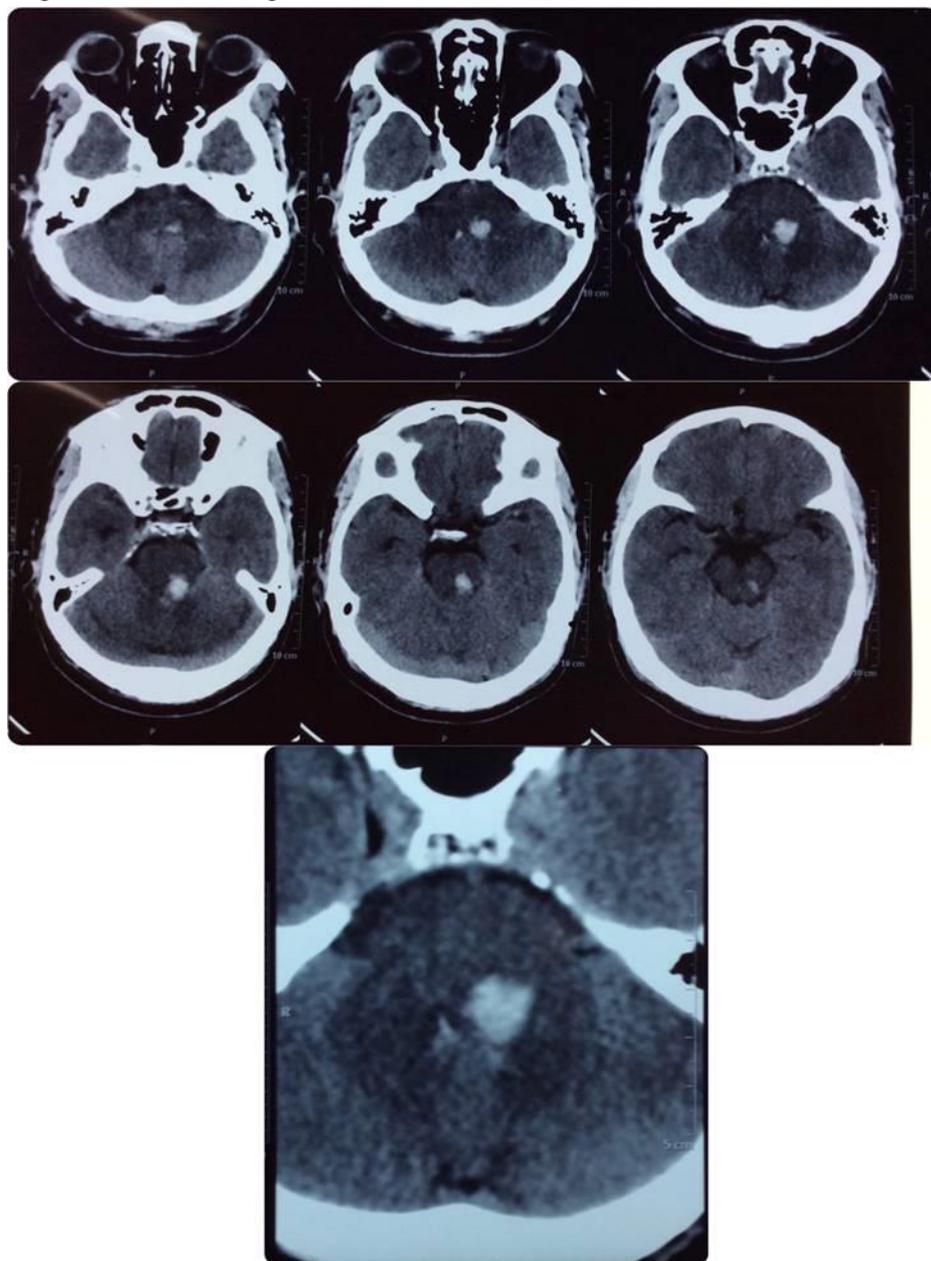


Figure – 2: CT brain plain showing intra cranial hemorrhage in the left side of the Pontine Tegmentum extending in to the 4th ventricle and left cerebellum.



On investigations - Hb-11g%, RBC count 4.2 mill/cumm, WBC count 7500 cells/cumm with Neutrophils - 80%, Lymphocytes - 24%, Eosinophils - 2%, Basophils - 0%, Monocytes - 2%, Platelet count was normal. ESR 1st hr - 20 mm, Blood Urea 20 mg%, Serum Creatinine - 0.6 mg, Complete Urine Examination (CUE) – normal, Random Blood Sugar - 103 mg/dl, Serum Electrolytes: Na – 143 mmol/L, K - 3.6 mEq/L, CL – 109 mEq/L. ECG revealed LVH.

Computed tomography (CT) scan brain showed intra cranial hemorrhage in the left side of the pontine tegmentum extending in to the 4th ventricle and left cerebellum (**Figure – 2**).

A diagnosis of Eight and a Half Syndrome associated with ipsilateral ataxia, due to pontine hemorrhage, hypertension was made.

The patient was treated with antihypertensives, anti-edema measures and nimodipine.

The drugs given were

Tab. Nicarda retard 10 mg 1 tid,

IV Mannitol 10% 100 ml tid for 3 days

Tab Nimodipine 30 mg tid for 6 weeks

Later on 30 mg tid for 3 months

Patient recovered completely after 2 months of therapy.

Discussion

The combined lesions of one and half syndrome and ipsilateral fascicular cranial VIIth nerve palsy is included under the extended spectrum of one and a half syndrome and is often labelled as eight and a half syndrome. Eggenberger [3] reported three cases of eight and a half syndrome in detail. The major structures involved in the horizontal eye movements are Para Median Pontine Reticular Formation (PPRF), abducens nucleus and Medial Longitudinal Fasciculus (MLF).

Pathophysiology

The supranuclear center for the control of horizontal conjugate eye movements is PPRF and is located near the ipsilateral abducens nucleus and ventral MLF and it receives inputs from the visual area of frontal lobe [4]. The excitatory neurons of the PPRF innervates the ipsilateral abducens nucleus and also controls the MLF, which in turn controls the contralateral oculomotor sub nucleus and subsequently the contralateral medial rectus, thus coordinating ipsilateral eye ball abduction and contralateral eye ball adduction resulting in conjugate movements of the eye [5]. Thus ipsilateral lesion of the PPRF results in horizontal gaze palsy on same side.

The abducens nucleus located in facial colliculus, has two distinct cell groups, the abducens motor neurons that innervate the lateral rectus muscle and the inter nuclear neurons whose axons cross the midline and ascend via the contralateral MLF to oculomotor sub nucleus and controls the medial rectus [6, 7]. Hence, Ipsilateral lesion of abducens nucleus also produce Ipsilateral horizontal gaze palsy.

One and a half syndrome

One and a half syndrome is a disorder of horizontal ocular movement characterized by lateral gaze palsy on looking towards the same side of lesion and a Inter Nuclear Ophthalmoplegia (INO) on looking to opposite direction. The location of the lesion is in the PPRF or VI nerve nucleus and MLF fibers crossing from the contra lateral VI nerve nucleus is also involved, causing INO.

The common causes of this syndrome are similar to those of INO (e.g., Multiple Sclerosis, CVA, hemorrhage or tumor in the lower pons). The other differential diagnosis are Myasthenia gravis, Fisher syndrome, Guillain-barre syndrome, Wernickes encephalopathy and frontoparietal infarct.

Clinical signs include the following

- Horizontal gaze palsy on looking towards the side of the lesion (“one”).
- INO on looking opposite to the side of the lesion (“half”).

Clinically characterized by an ipsilateral adduction deficit (partial or complete) with a contralateral, dissociated, horizontal abducting saccade on attempted gaze to contralateral side. As a result, ipsilateral eye has no horizontal movement, and the only lateral ocular movement possible is abduction and nystagmus of contralateral eye.

Herring’s law of equal innervation has been hypothesized as a possible reason for the dissociated contralateral gaze evoked nystagmus in the abducting eye. The increased innervation to the underacting adducting muscle would result in enhanced stimulus to the contralateral abducting muscle [8]. Associated signs include skew deviation, gaze – invoked nystagmus on vertical gaze, and an intermitted exotropia of the eye contralateral to the lesion is often present, which would be more pronounced on contralateral gaze. If the ipsilateral abducens, rather than the PPRF is involved, the

contralateral adduction deficit may be less pronounced, but an ipsilateral facial palsy may now manifest [8]. Vertical ocular movements and convergence are usually intact. MRI usually detects tumors, myelinolysis, and vascular malformation, but only sometimes detects stroke and multiple sclerosis.

Treatment is usually according to the etiology like, Stroke: supportive care, Tumor: search for primary site and treat accordingly, Multiple sclerosis: immunomodulatory agents, Fisher syndrome: plasmapheresis or intravenous immunoglobulin, Wenicke's encephalopathy: thiamine.

INO

INO is an eye movement disorder, where there is an impaired adduction on the side of the lesion involving the MLF, with dissociated nystagmus of the abducting eye [9]. This ophthalmoplegia is due to damage to the interneuron between the two nuclei of cranial nerves (CN) VI and (CN) III (internuclear). This interneuron is called the MLF. The MLF can be damaged by any lesion (e.g., demyelinating, ischemic, neoplastic, inflammatory) in the pons or midbrain. MLF is a myelinated nerve tract connecting the CN III of ipsilateral side with the PPRF and Cranial nerve VI of the opposite side.

Associated syndromes

Eight and a half syndrome

The combined lesion horizontal gaze palsy along with INO constitutes one and a half syndrome and additional involvement of LMN facial palsy as eight and a half syndrome and the anatomical localization is in the Ipsilateral dorsal tegmentum of the caudal pons [10].

Half and a Half syndrome

A syndrome consists of an INO in one eye combined with an ipsilateral CN VI fascicular involvement with sparing of the sixth nerve nucleus. Hence, there is "half" of a horizontal gaze palsy (INO) plus an additional "half" (abduction deficit from CN VI fascicular palsy).

WEBINO (Wall-Eyed Bilateral Internuclear Ophthalmoplegia)

Wall-Eyed Bilateral Internuclear Ophthalmoplegia exists when there is bilateral damage to the MLF, this damage causes primary position exotropia of bilateral eyes. The most common etiology is infarction of the mid-brain in older individuals and demyelinating diseases in younger patients, the exotropia is likely decompensation of fusional mechanisms and is not present in every case of bilateral INO [11].

WEMINO (Wall-Eyed Monocular Internuclear Ophthalmoplegia)

Patients with a unilateral MLF lesion (monocular INO) have a primary position exotropia. This is a less common variant of INO, similar to the WEBINO as described above [12].

Posterior INO (Lutz)

This syndrome is a rare ophthalmoplegia, either bilateral or unilateral that exhibits contralateral adducting eye (rather than abducting eye) nystagmus with abduction restriction on examination. It is the reverse of typical INO. The lesion localisation is not consistent, likely due to VIth cranial nerve pre-nuclear input asymmetry [13].

The etiology for these syndromes are infarcts/hemorrhage in older people, demyelinating disorders, brain stem tumors, inflammatory (e.g., sarcoidosis, Behcet's disease, lupus) or infectious (e.g., Lyme disease, cryptococcosis) and AV malformations [14, 15]. Rossini, et al. described a possible nine syndrome which is eight and a half syndrome with hemiparesis and hemihypesthesia due to additional involvement of corticospinal tract and medial lemniscus by lacunar pontine infarction [18].

Lateral Tegmental Brainstem Hematomas

Lateral tegmental brainstem hematomas usually originate from vessels penetrating in to the brainstem from long circumferential branches. Small hematomas remain confined to the lateral tegmentum, while larger lesions spread across to opposite side and destroy entire tegmentum.

Neurological examination reveals a predominantly unilateral tegmental lesion with variable degrees of basilar involvement [19]. Oculomotor abnormalities, especially the “one - and-a -half syndrome”, horizontal gaze palsy, INO, partial involvement of vertical eye movements, ocular bobbing, have been described.

The tegmental location of the spinothalamic tract makes sensory symptoms common. Ataxia, either unilateral or bilateral, may also accompany the oculomotor signs. Facial numbness, hemiparesis have also been noted. Rare cases of patients with Cheyne-stokes respirations, and action tremor have been described. The most distinctive and diagnostic of lateral tegmental pontine hematomas are the oculomotor abnormalities, which include Ipsilateral conjugate gaze paresis, Ipsilateral INO, or a one-and-a half syndrome.

In the present case, the patient had clinical features of eight and a half syndrome due to caudal pontine tegmental bleeding and additional finding of left cerebellar gait ataxia was due to extension of bleeding into the cerebellum.

Prognosis

The prognosis of One and a half syndrome or INO is based on treatment of the underlying etiology. Ischemic and demyelinating cases typically recover. Patients with WEBINO, WEMINO, pontine paralytic or non- paralytic exotropia with residual primary position symptomatic deviation that do not recover may benefit from patching, prism, or strabismus surgery [20].

Conclusion

There are clinical variants in the eight and a half syndrome with clinical features depending on the adjacent structure involvement based on the extent of lesion from pontine tegmentum to various parts such as midbrain, medulla and cerebellum but the basic pathology of recognizing this syndrome will help to localize

the lesion to Ipsilateral pontine tegmentum. Here we are presenting a rare case of Eight and a Half Syndrome associated with ipsilateral ataxia, due to pontine hemorrhage with hypertension.

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