International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Eight-and-A-Half Syndrome: A Rare Case Report

Dr. Dhruvi Alpeshkumar Brahmbhatt[1], Dr. Ashish Sindhi[2]

[1]2nd Year Medicine Resident, Narendra Modi Medical College and L.G. Hospital, Ahmedabad, India
Email: *dhruviabarot[at]gmail.com*

[2]2nd Year Medicine Resident, Narendra Modi Medical College and L.G. Hospital, Ahmedabad, India

Abstract: ***Background: 1) Eight-and-a-half syndrome describes a constellation of symptoms that occur due a lesion involving the abducens (CN) nucleus, the fascicular portion of the facial (CN VII) nerve and the medial longitudinal fasciculus (MLF). 2) It results in a combination of horizontal gaze palsy, facial nerve palsy, and internuclear ophthalmoplegia (INO). 3) It is usually caused by demyelination, infarct, tumors and occasionally by haemorrhage. Case Presentation: 1) A 58 year old female known to have hypertension presented with deviation of angle of mouth towards right side, inability to close left eye lid and slurring of speech since 3 days. 2) On examination of extraocular movement, left conjugate horizontal gaze palsy with limited right abduction and horizontal nystagmus of right eye was seen which were suggestive of left sided One-and-a-half syndrome. 3) Cranial nerve examination showed left lower motor neuron facial nerve palsy. Rest neurological examination was normal. 4) Magnetic resonance imaging of brain showed left frontal, thalamic and pontine small foci of acute infarcts and left thalamic chronic haemorrhage. 5) Patient was treated with antihypertensives, antiplatelets and artificial eye drops. Conclusion: The rare diagnosis of Eight-and-a-half syndrome requires high degree of clinical suspicion when comorbid eye movement limitation and peripheral facial paralysis are seen followed by thorough clinical examination for identifying precise location of the lesion and to determine proper treatment regime for the patient.***

Keywords: Eight-and-a-half, abducens nucleus, facial nerve, internuclear ophthalmoplegia

Case Report

1. Introduction

- Eight-and-a-half syndrome first described by Eggenberger in 1998, is a rare pontine neuro ophthalmological syndrome consisting of conjugate horizontal gaze palsy, ipsilateral internuclear ophthalmoplegia and ipsilateral lower motor neuron-like facial palsy.
- This is caused by a lesion that affects the ipsilateral paramedian pontine reticular formation (PPRF) or the abducens (CN VI) nucleus (responsible for the horizontal gaze palsy) and the ipsilateral medial longitudinal fasciculus (MLF) (responsible for INO – failed adduction of the ipsilateral eye)

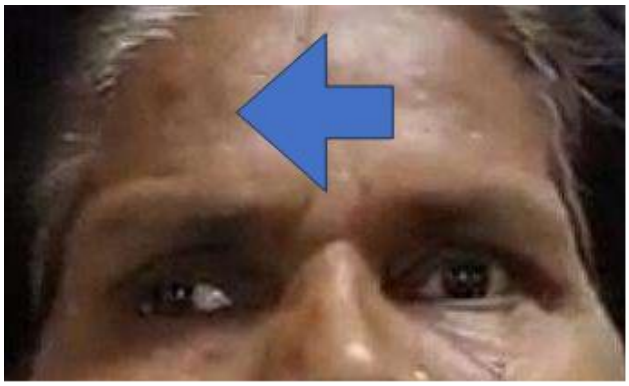
2. Case Description

- A 58 year old female known to have hypertension presented with deviation of angle of mouth towards right side, inability to close left eye lid and slurring of speech since 3 days.
- On admission patient's blood pressure was 180/96mmHg, rest vitals were normal.
- General physical examination was unremarkable.
- On examination of extraocular movement, left conjugate horizontal gaze palsy with limited right abduction and horizontal nystagmus of right eye was seen; these findings are suggestive of left Internuclear ophthalmoplegia. Left eye abduction weakness was much more than the adduction weakness resulting in adducted eye in neutral position this is because of involvement of abducens nucleus. Cranial nerve examination showed left lower motor neuron facial nerve palsy. Rest neurological examination was normal.

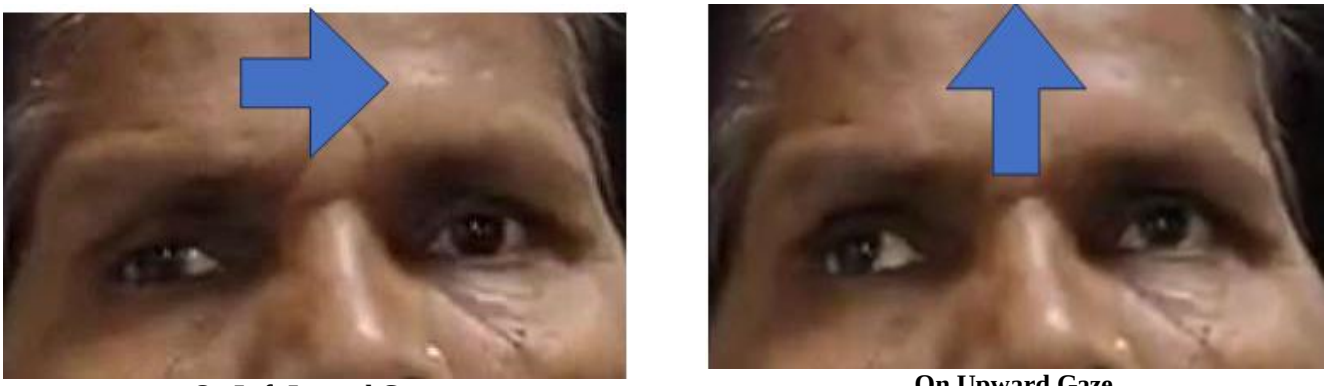
3. Investigations

HB: 12.3 TC: 9900 APC: 3.21
UREA: 36CREAT:0.79NA:139
K: 4.10 SGPT: 14 ALP: 137
TB/DB:1.1/0.4 CPKMB: 23
TROPI: 0.01

2DECHO:
ReducedLVcompliance
Ef: 60%
TrivialMR Mild TR
No PAH
Mildsclerosed



On Right Lateral Gaze



On Left Lateral Gaze

On Upward Gaze

Figure 1

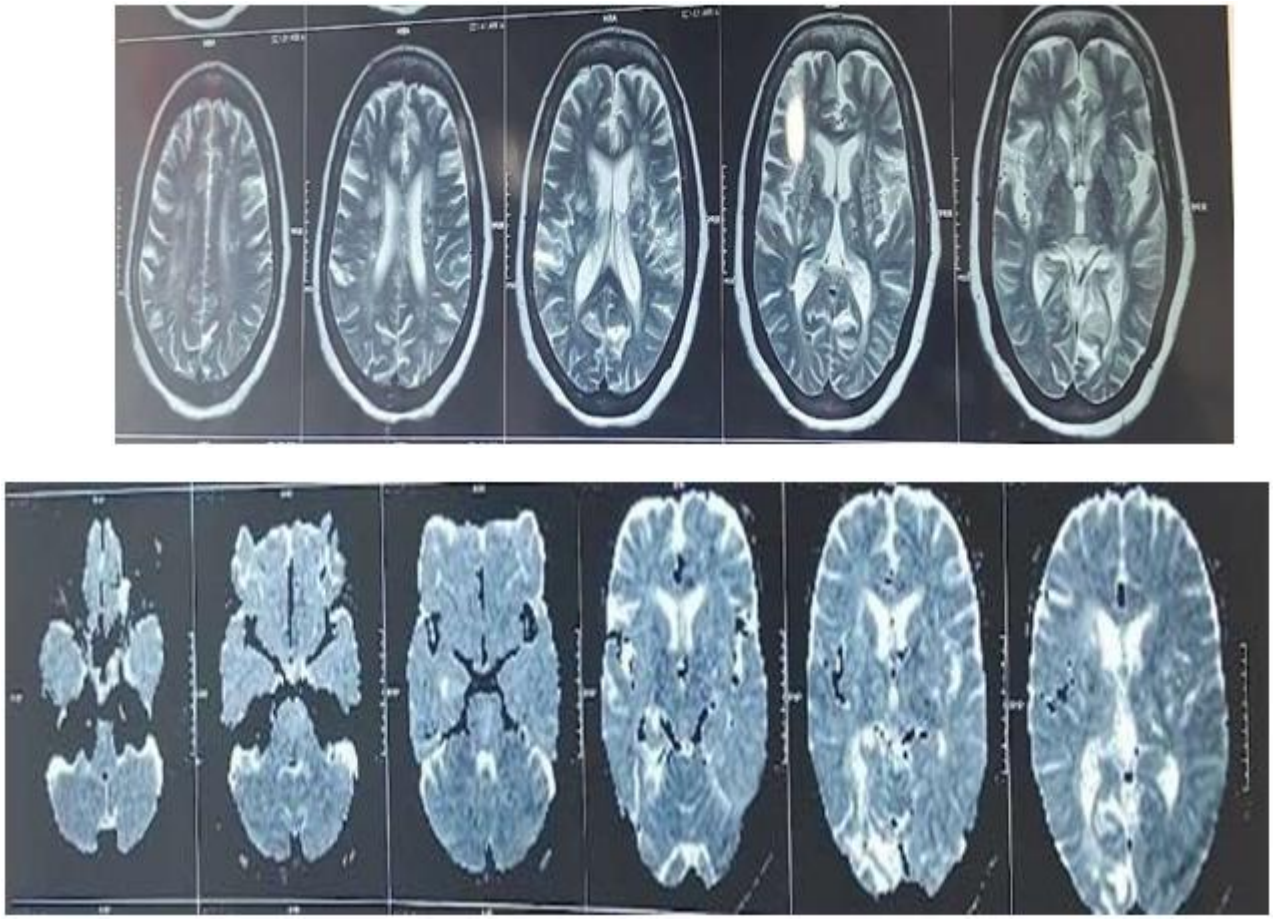


Figure 2: The MR findings show

- Left frontal, thalamic and pontine small foci of acute infarcts.
- Left thalamic chronic hemorrhage. Rightoccipitalgliosis.
- Multiple discrete & confluent chronic ischemic changes in bilateral fronto-parietal sub-cortical & peri-ventricular
- White matter and pons.
- Age-related cerebral and cerebell aratrophic changes.

Treatment

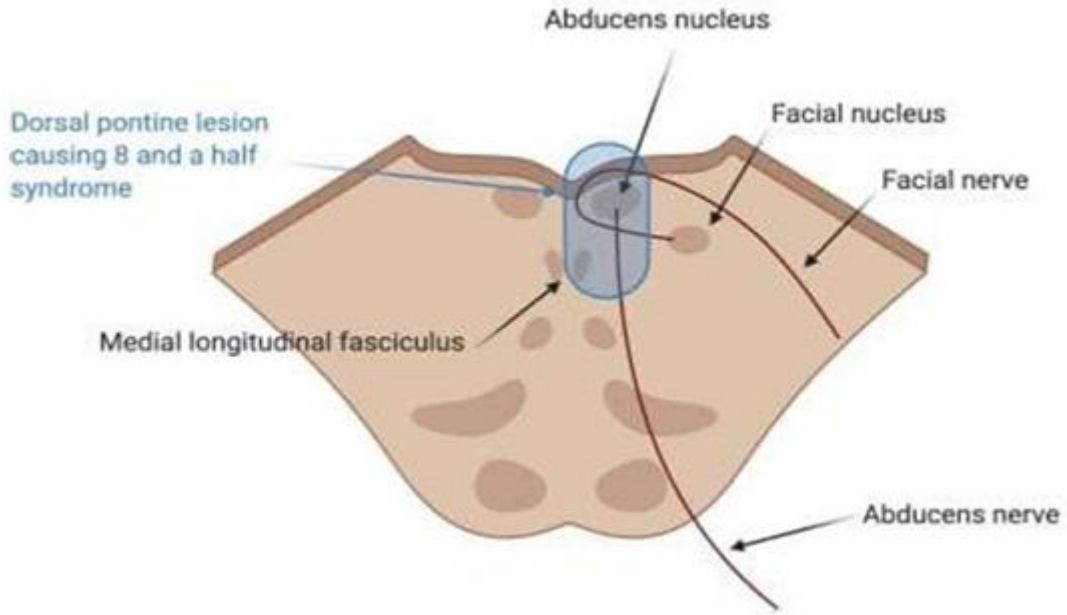
Patient was treated with antihypertensive, antiplatelets, dyslipidemic drugs and artificial eye drops.

4. Discussion

- One-and-a-half syndrome (OHS) refers to horizontal gaze palsy with internuclear ophthalmoplegia due to a lesion of the paramedian pontine reticular formation (PPRF) and medial longitudinal fasciculus.
- "Eight-and-a-half" syndrome is "one-and-a-half" syndrome (conjugated horizontal gaze palsy and internuclear ophthalmoplegia) plus ipsilateral facial nerve (CN VII) palsy.
- The facial nucleus lies adjacent to the PPRF and gives rise to cranial nerve VII (CN7), which courses through the PPRF before encircling the abducens nucleus and exiting the brainstem.
- PPRF receives impulse from contralateral frontal eye field (FEF) and ipsilateral parietal cortex, acting as the last supranuclear relay involved in conjugate horizontal eye movement. The impulse is then relayed to ipsilateral abducens nucleus and via the MLF to contralateral oculomotor nucleus, causing contraction of ipsilateral

lateral rectus and contralateral medial rectus muscle, resulting in horizontal gaze.

- This rare condition, particularly when isolated, is caused by circumscribed lesions of the pontine tegmentum as a result of infarction, hemorrhage , demylination (Multiple Sclerosis),gliomas and cystic lesions.



5. Conclusion

- Diagnosing eight and half syndrome requires high degree of clinical suspicion and thorough clinical examination for identifying precise location of the lesion.
- As most of the patients presents with an LMN type of facial nerve palsy as this is the most visible component of the condition. In addition, patient's rarely complain of diplopia in horizontal gaze palsies, and hence, the diagnosis may be missed. So, a mandatory extraocular movement testing should be done in all patients with facial nerve palsy to pick up associated gaze palsies. The presence of one-and-a-half syndrome associated with LMN type of facial nerve palsy will help the physician to accurately pinpoint the lesion at the ipsilateral pontine level.

References

[1] https://eyewiki.org/Eight-and-a-Half_Syndrome
[2] https://eyewiki.org/File:Pons.png
[3] Eggenberger E. Eight-and-a-half syndrome: one-and-a-half syndrome plus cranial nerve VII palsy. J Neuroophthalmol. 1998; 18(2):114–6.
[4] Kakar P, Brown Z, Banerjee S. Eight-and-a-half syndrome: an unusual presentation of brainstem infarction. *QJM*. 2013 Mar; 106(3):273–276. doi: 10.1093/qjmed/hcr187. Available from: http://dx.doi.org/10.1093/qjmed/hcr187.
[5] https://jmedicalcasereports.biomedcentral.com/articles/10.1186/s13256-022-03699-8
[6] https://jbcahs.sbvjournals.com/abstractArticleContentBrowse/JBCAHS/25035/JPJ/fullText