Pakistan Journal of Neurological Sciences (PJNS)

Volume 10 | Issue 1 Article 8

3-2015

Eight – and - half syndrome: a rare neuro-ophthalmic syndrome "Possible Nine Syndrome"

Muslim Ali Lakhiar

Shaheen Ahmed Mughal

EIGHT -AND-HALF SYNDROME: A RARE NEURO-OPHTHALMIC SYNDROME "POSSIBLE NINE SYNDROME"

Dr. Muslim Ali Lakhiar, Dr. Shaheen Ahmed Mughal

Correspondence to: *Dr. Muslim Ali Lakhiar, Assistant Professor Neurology. PUMHS Nawabshah-SBA. Email: drmuslimali@yahoo.com*
Date of Submission: *November 29, 2014,* ***Date of Revision:*** *December 28, 2014,* ***Date of Acceptance:*** *January 3, 2015*

ABSTRACT

"Eight-and-half" syndrome is "one-and-a-half" syndrome characterized by conjugated horizontal gaze palsy and inter-nuclearopthalmoplegia plus ipsilateral fascicular seventh nerve palsy. We report a case of 50 year old woman who presented with right eight and half syndrome together with contralateral hemiparesis and hemihypesthesia. Non contrast CT scan brain showed hyperdensesignal on right pontine region. MRI brain plain revealed hemorrhagicpontine-tegmentuminfarction as well as corticospinal tract and medial meniscus. This spectrum of presentation suggests a possible "Nine syndrome" as a novel neuropthalmicpontine syndrome. The first of its kind ever reported from Pakistan

Key words: infarction, hemorrhage, eight-and-halfSyndrome, nine syndrome.

INTRODUCTION

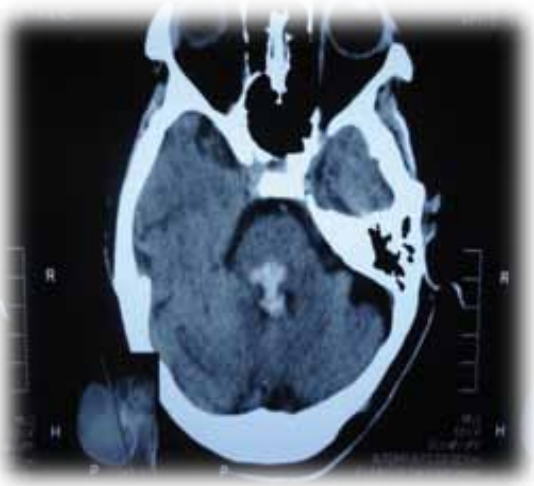
The "one and ahalf " syndrome, first reported and named byMiller Fisher in 1967[1], is a clinical syndrome of horizontal gazepalsy to one side along with ipsilateral inter-nuclearopthalmoplegia (INO) resulting in loss of allhorizontal movements in the ipsilateral eye, except-abduction of the contralateral eye. This syndrome is due to a unilateral lesion in the lower part of the dorsal pontinetegmentum, affecting the ipsilateralparamedi-anpontine reticular formation (PPRF)which serves as the conjugate gaze center for horizontal eye movements, abducensnucleus and internuclear fibers of the ispsilateral medial longitudinal fasciculus (MLF). It is caused by circumscribed lesions of the pontinetegmentum as a result of infarction, hemorrhage, demyelination (Multiple Sclerosis), gliomas, and cystic lesions.It is one of the most localizing brainstem syndromes. Involvement of facial nervewith "one and a half" syndrome has been termed as "eight and a half" syndrome by Eggen-Berger [2] whofirst reported three such cases. Keeping this in view we report another rare case of "Eight and a half" syndromewith additional features of contralateral hemi-paresis & hemihypoesthesia as a possible "NINE Syndrome".

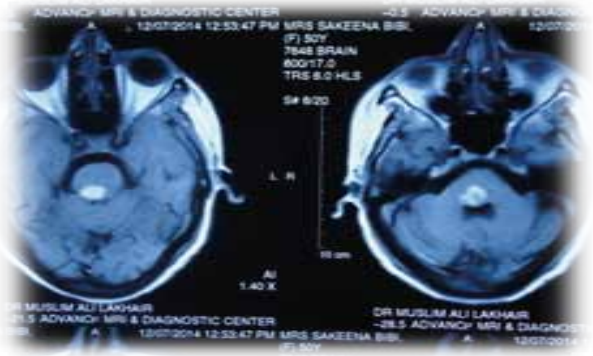
CASE PRESENTATION

A 50 year old female, right handed, known case of Hypertension and Diabetes mellitus admitted through casualty in Medical ward with sudden onset altered level of consciousness, left sided weakness, facial asymmetry, diplopia in horizontal gaze and two episodes of non projectile vomiting. At the time of presentation her blood pressure was 200/100mmHg, Pulse 90 b/min, respiratory rate 17 breaths/min and Tem.98F. On neurologic examination she had a Glasgow coma scale 11/15, Pupils 4mm and reactive bilaterally, fundoscopy normal, oculomotor examination revealed conjugated horizontal gaze paresis. Corneal reflex showed blink response on left side only, she had a right seventh nerve palsy which was lower motor neuron type and rest of cranial nerve were normal. On motor system examination bulk was normal, tone decreased on left side in upper and lower limb, power was 0/5 in upper limb and 3/5 in lower limb on left side while reflexes were 3+ and left planter was upgoing. The Respiratory system, cardiovascular and abdomen were all normal ABC was maintained & patient was started on i/v 3rd generation cephalosporin, tablet enalapril 10mg and regular insulin according to sliding scale while RBS done at presentation was 234mg/dl, meanwhile base-line investigations sent. On second day of admission her blood pressure dropped to 160/90mmgh and GCS became 15/15. However oculomotor examination revealed conjugated horizontal gaze paresis except left sided nystagmus on abduction while from primary position vertical eye movements were normal, power was same. Cerebellar signs normal. Pinprick, temperature, vibration &joint position sense were diminished on left side. Laboratory investigations showed a raised WBC count 13000/mm3 with platelet count of 373000, while blood urea, creatinine, HbsAg, Anti HCV, LFTs, electrolytes, APPT, PT-INR, Bleeding time and clotting time were normal. Urine detailed report showed pus

cells. ECG was unremarkable and Hemoglobin A1c was 7.3% (4.0-7.0%). Non contrast CT scan brain showed hyperdensesignal on right pontine tegmental region. Computed Tomographic Angiography did not reveal any aneurysm or vascular malformation, MRI brain plain done one week later revealed hyperintense signals over right pontinetegmentum on T1,T2 and flair imaging suggesting subacute hemorrhagic pontine infarction involvingcorticospinal tract and medial meniscus as well. MRA was unremarkable. Over a period of 1 week the condition of patient gradually improved in terms of power which became 1/5 in upper limb and 4/5 in lower limbbut her diplopia in horizontal gaze remained same. Patient was discharge on antihypertensive and oral hypoglycemic agents to follow up in Neurology OPD.

Figure -01 showing eye movement abnormality.





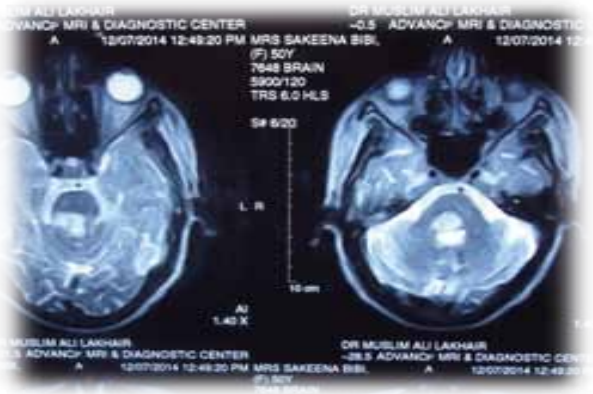


Figure-02 Non contrast CT scan showing hyperdense signal at right pontinetegmentum extending to 4th Ventricle&MRI Brain Plain showing Hyperintense signals on T1WI, T2WI and Flair imaging.

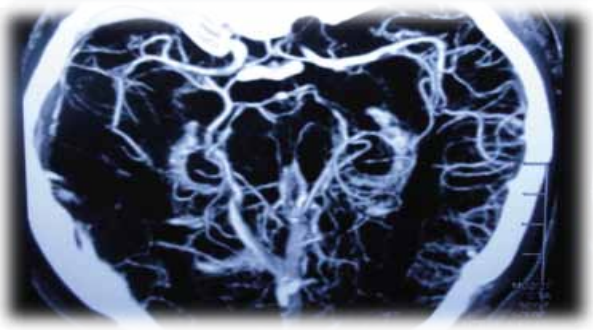


Figure-03 Magnetic Resonance Angiography (MRA) & CT Angiography Normal

DISCUSSION

One-and-a-half syndrome (OHS) refers to horizontal gaze palsy (one) with intranuclearopthalmoplegia (half) due to a lesion of the paramedianpontine reticular formation (PPRF) and medial longitudinal fasciculus. [1]. When one-and-a-half syndrome is associated with other cranial nerve involvement, the number of the cranial nerve is added to one-and-a-half to describe the clinical syndrome like eight and half syndrome [2,3,4], Nine syndrome [5], thirteen and half syndrome[6], Fifteen and half syndrome [7], sixteen and half syndrome [8,9]. Hence, "Eight-and-a-half" syndrome is "one-and-a-half" syndrome (conjugated horizontal gaze palsy and internuclearopthalmoplegia) plus ipsilateral fascicular cranial nerve seventh palsy.Eight-and-a-half syndrome is caused by a lesion in the dorsal tegmentum of the caudal pons involving the PPRF or abducens nucleus and the medial longitudinal fasciculus (MLF), as well as the nucleus and fasciculus of the facial nerve [3]. The facial nucleus lies adjacent to the PPRF and gives rise to cranial nerve VII (CN7) which courses through the PPRF before circling the abducens nucleus and exiting the brainstem. This rare condition, particularly when isolated, is caused by circumscribed lesions of the pontinetegmentumas a result of infarction, hemorrhage, demyelination (Multiple Sclerosis), gliomas, and cystic lesions. Our case presented with eight and a half syndrome associated with contralateral hemiparesis and hemihypesthesia, Almost similar clinicalentity has been termed as "Nine syndrome" previously reported by Rosni at al [5] resulting from lacunar pontine infarction. Our patient was younger who developedhemorrhagic pontinetegmentum infarction as a result of high blood pressure. Pons is one of the most common sites for hemorrhagic stroke apart from putamen, thalamus and cerebellum.No such clinical entity has been reported yet in Pakistan.This is an addition in the spectrum of brainstem syndromes which not only have clinical importance but also help anatomic localization from student's point of view as well in order to have appropriate imaging.

CONCLUSION

Recognition of one-and-a-half plus syndrome at the bedside is of importance for precise localization and also for planning appropriate investigations to determine the possible cause.

REFERENCES

1. Fisher CM. Some neuro-ophthalmological observations. J NeurolNeurosurg Psychiatry. 196 7;30:38 3-92.
2. Eggenberger E. Eight-and-a-Half Syndrome: One-and-a-Half Syndrome plus Cranial Nerve VII Palsy. J Neuro-ophthalmology. 1998;18:114-116.
3. SarwalA, Garewal M, Sahota S, Sivaraman M. Eight-and-a-half syndrome. Journal of Neuroimaging. July 2009;19(3):288-90.
4. Nandhagopal R Krishnamoorthy SG. Eight-and-a-half syndrome. J NeurolNeurosurg Psychiatr y. April 2006;77(4): 463.
5. Rosini F, Pretegiani E, Guideri F, Cerase A, Rufa A, Eight and a Half Syndrome with Hemiparesis and Hemihypesthesia: The Nine Syndrome? J Stroke-Cerebrovas Dis. November 2013;22(8):637–638.
6. Rath A, Jawatkar J, Guruprasd H, Saxena A. B, Murthy J. M. K.One-and-a-half Plus Syndromes: Report of Three Cases .Annals of Indian Academy of Neurology; 2007 Supplement 2, Vol. 10, p6
7. Bae JS, Song HK.One-and-a-Half Syndrome with Facial Diplegia: The 15 1/2 Syndrome?. Journal of Neuro-opthmology. March 2005;25(1)55-53
8. Cummins G, O'Hare A, Dunne R, et al. "Sixteen and a half": a novel pontineneuroophthalmological syndrome. J Neurol. July 2011;258(7):1347-8.
9. Singh et al / "Sixteen and a half": a rare neurological syndrome. Internet Journal of Medical Update. July 2012;7(2):51-53

Conflict of Interest: Author declares no conflict of interest.

Funding Disclosure: Nil

Author's contribution:

Dr. Muslim Lakhiar: Study concept and design, protocol writing, data collection, data analysis, manuscript anuscriwriting, mpt review

Dr. Shaheen Mughal: Study concept and design, data collection, data collection, data analysis, manuscript writing, manuscript review.