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

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ABSTRACT

“Eight-and-half” syndrome is “one-and-a-half” syndrome characterized by conjugated horizontal gaze palsy and internuclear ophthalmoplegia 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 hyperdense signal on right pontine region. MRI brain plain revealed hemorrhagic pontine tegmentum infarction as well as corticospinal tract and medial meniscus. This spectrum of presentation suggests a possible “Nine syndrome” as a novel neurophthalmic pontine syndrome. The first of its kind ever reported from Pakistan

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

INTRODUCTION

The “one and a half” syndrome, first reported and named by Miller Fisher in 1967^[1], is a clinical syndrome of horizontal gaze palsy to one side along with ipsilateral internuclear ophthalmoplegia (INO) resulting in loss of all horizontal 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 pontine tegmentum, affecting the ipsilateral paramedian pontine reticular formation (PPRF) which serves as the conjugate gaze center for horizontal eye movements, abducens nucleus and internuclear fibers of the ipsilateral medial longitudinal fasciculus (MLF). It is caused by circumscribed lesions of the pontine tegmentum 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 nerve with “one and a half” syndrome has been termed as “eight and a half” syndrome by Eggen-Berger^[2] who first reported three such cases. Keeping this in view we report another rare case of “Eight and a half” syndrome with additional features of contralateral hemiparesis & 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 Temp. 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 baseline investigations sent. On second day of admission her blood pressure dropped to 160/90mmHg 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 hyperdense signal 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 pontine tegmentum on T1, T2 and flair imaging suggesting subacute hemorrhagic pontine infarction involving corticospinal 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 limb but 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.

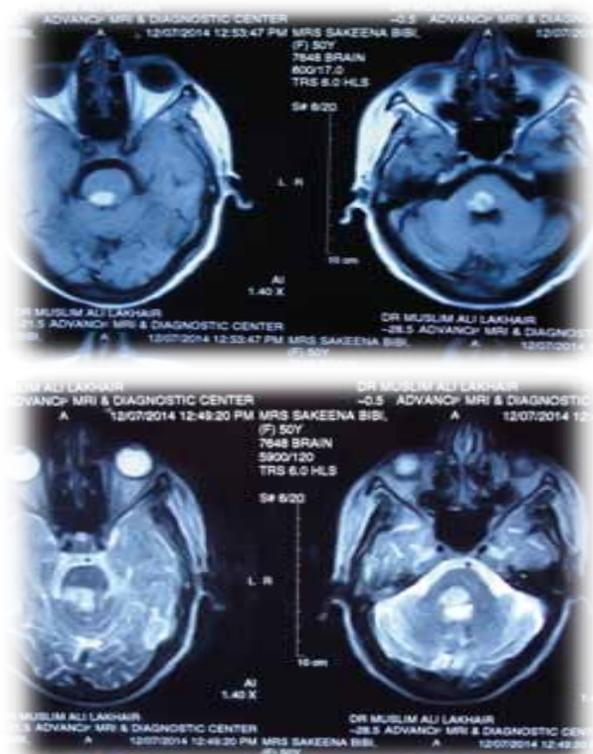


Figure-02 Non contrast CT scan showing hyperdense signal at right pontine tegmentum extending to 4th Ventricle & MRI Brain Plain showing Hyperintense signals on T1WI, T2WI and Flair imaging.

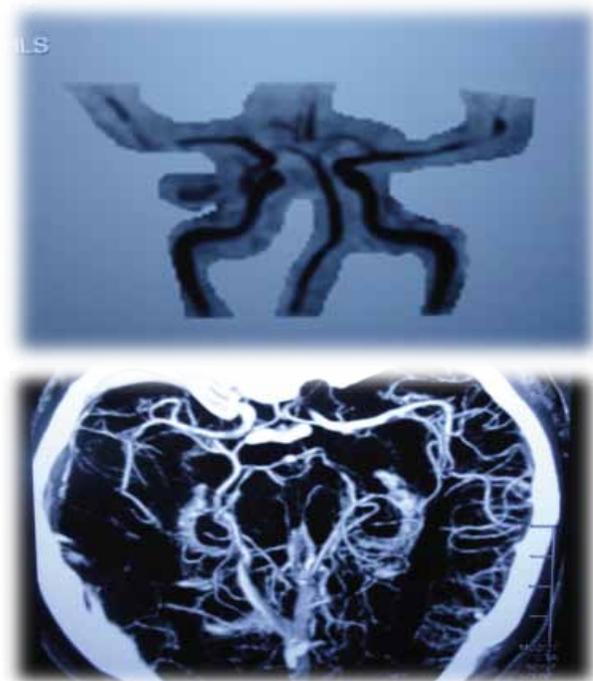


Figure-03 Magnetic Resonance Angiography (MRA) & CT Angiography Normal

DISCUSSION

One-and-a-half syndrome (OHS) refers to horizontal gaze palsy (one) with intranuclear ophthalmoplegia (half) due to a lesion of the paramedian pontine 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 internuclear ophthalmoplegia) 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 pontine tegmentum as 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 clinical entity has been termed as "Nine syndrome" previously reported by Rosni et al [5] resulting from lacunar pontine infarction. Our patient was younger who developed hemorrhagic pontine tegmentum 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.

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Author's contribution:

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

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