Stroke Syndromes: A Case of an Eight and a Half Syndrome

Karlo Angelo F. Camagay, MD[1] and Maria Socorro F. Sarfati, MD, FPNA[2]

ABSTRACT

Introduction

Eight and a Half syndrome: a combination of ipsilateral cranial nerve seven palsy plus one and a half syndrome is rare**.** Exact prevalence of the syndrome has not been reported as of yet. This syndrome is mostly attributed to a vascular etiology such as a pontine tegmental infarction.

Objective

To present a rare case of a stroke syndrome : eight and a half syndrome (peripheral cranial nerve seven palsy plus a one and an half syndrome) in an adult male. To present the importance of its early clinical recognition in correlation of radiologic imaging, and management.

Case Report

This is a case of a sixty-two year old male, who had a one day history of sudden double vision. Cranial nerve examination revealed a frozen right eye; unable to perform any movement on horizontal gaze, and with right sided facial asymmetry. He was hypertensive for more than ten years. Left eye was exotropic, with no adduction. Right eye was frozen on horizontal gaze, and primary gaze was at midline. Right sided peripheral facial palsy was seen on examination. Cranial non-contrast magnetic resonance imaging with time of flight was done revealing an infarct in the right posterior pontine area, and a narrow right vertebral artery due to a probable occlusion. Patient was started on antiplatelet cilostazol 100mg/tab 1 tablet twice daily. Atorvastatin 40mg/tab 1 tablet was given. Anti-hypertensives were started on his fourth hospital day. Smoking cessation, dietary modifications, and compliance to medications were emphasized prior to discharge.

Discussion

Here we have a stroke syndrome presenting as an ipsilateral lower motor neuron: seventh nerve palsy, and an ipsilateral horizontal gaze palsy with internuclear ophthalmoplegia of the contralateral eye (failure of adduction) termed as CN VII, + 1 ½ syndrome or Eight and a Half Syndrome. This is caused by a lesion involving the paramedian pontine reticular formation (PPRF) which sends signals towards the ipsilateral abducens nerve and contralateral medial longitudinal fasciculus. These structures lie in close proximity to the nucleus and intraaxial fascicles of cranial nerve VII manifesting as facial weakness of the ipsilateral side to the lesion. An occlusion in the tip of the paramedian pontine artery, a branch of the basilar artery, is the most common etiology.

Keywords: *Eight and a Half syndrome, Posterior circulation infarcts*

From the Department of Neurology, Baguio General Hospital
[1]Adult Neurology Resident -in-Charge, [2]Research Consultant

INTRODUCTION

Eight and a Half syndrome: a combination of ipsilateral cranial nerve seven palsy plus one and a half syndrome is rare.[1,2,3] With the advent of the use of magnetic resonance imaging in correlating the localization of focal brainstem deficits, cases began to be reported.[4,5] Exact prevalence of the syndrome has not been reported as of yet. This syndrome is mostly attributed to a vascular etiology such as a pontine tegmental infarction.[5,6]

OBJECTIVE

To present a rare case of a stroke syndrome: eight and a half syndrome (peripheral cranial nerve seven palsy plus a one and a half syndrome) in an adult male. To present the importance of its early clinical recognition in correlation of radiologic imaging, and management.

CASE PRESENTATION

This is a case of a sixty-two year old male, right handed who had a one day history of sudden double vision when patient looks to his left, with noted frozen right eye; unable to perform any movement on horizontal gaze, and with right sided facial asymmetry. There were no noted complaints of dizziness, headache, vomiting, or weakness of extremities. Patient's blood pressure during this time was 180/100 mmHg. Due to the elevated blood pressure reading and evident neurological deficits, patient sought consult in our institution. He was hypertensive for more than ten years, taking losartan 50mg/tab one tablet daily, but was non-compliant. No other co-morbidities were extracted from the patient. He has a strong family history of hypertension, as both his parents were hypertensive along with all his siblings, and he was a forty pack year smoker as well.

Neurologic examination showed intact higher cortical functions with a mini mental status exam 30/30 (normal). Both eyes were non-ptotic, Left eye was exotropic, confirmed by an asymmetrical Hirschberg test, with no adduction and with nystagmus upon abduction. Right eye was frozen on horizontal gaze, and primary gaze was at midline. Pupils were equally reactive to direct and consensual reflex. Right sided peripheral facial palsy was seen on examination. Other cranial nerve findings were unremarkable. No motor weakness and no sensory deficits were elicited. Deep tendon reflexes were normal and cerebellar signs and meningeal signs were absent.

Cranial non-contrast magnetic resonance imaging with time of flight was done revealing an infarct in the right posterior pontine area, and a narrow right vertebral artery due to a probable occlusion. Patient was started on antiplatelet cilostazol 100mg/tab 1 tablet twice daily. Atorvastatin 40mg/tab 1 tablet was given. Anti-hypertensives was started on his fourth hospital day. Smoking cessation, dietary modifications, and compliance to medications were emphasized prior to discharge. He was then discharged improved.

DISCUSSION

Presented is a hypertensive smoker with noted end organ damage which was manifested by a brainstem infarct. Posterior circulation strokes account for 20-25% of ischemic strokes, with diplopia or double vision as a common presenting symptom [7]. Here we have a stroke syndrome presenting as an ipsilateral lower motor neuron: seventh nerve palsy, and an ipsilateral horizontal gaze palsy with internuclear ophthalmoplegia of the contralateral eye (failure of adduction) termed as CN VII, + 1 ½ syndrome or Eight and a Half Syndrome.[1,2,5,6]

This is caused by a lesion involving the paramedian pontine reticular formation (PPRF) which sends signals towards the ipsilateral abducens nerve (CN VI) and

Figure 1. Image on left demonstrates a peripheral right facial palsy. Images on the right from top to bottom illustrates eye movements, on primary gaze, right horizontal gaze and left horizontal gaze respectively. Frozen right eye, and failure of adduction of the left eye.

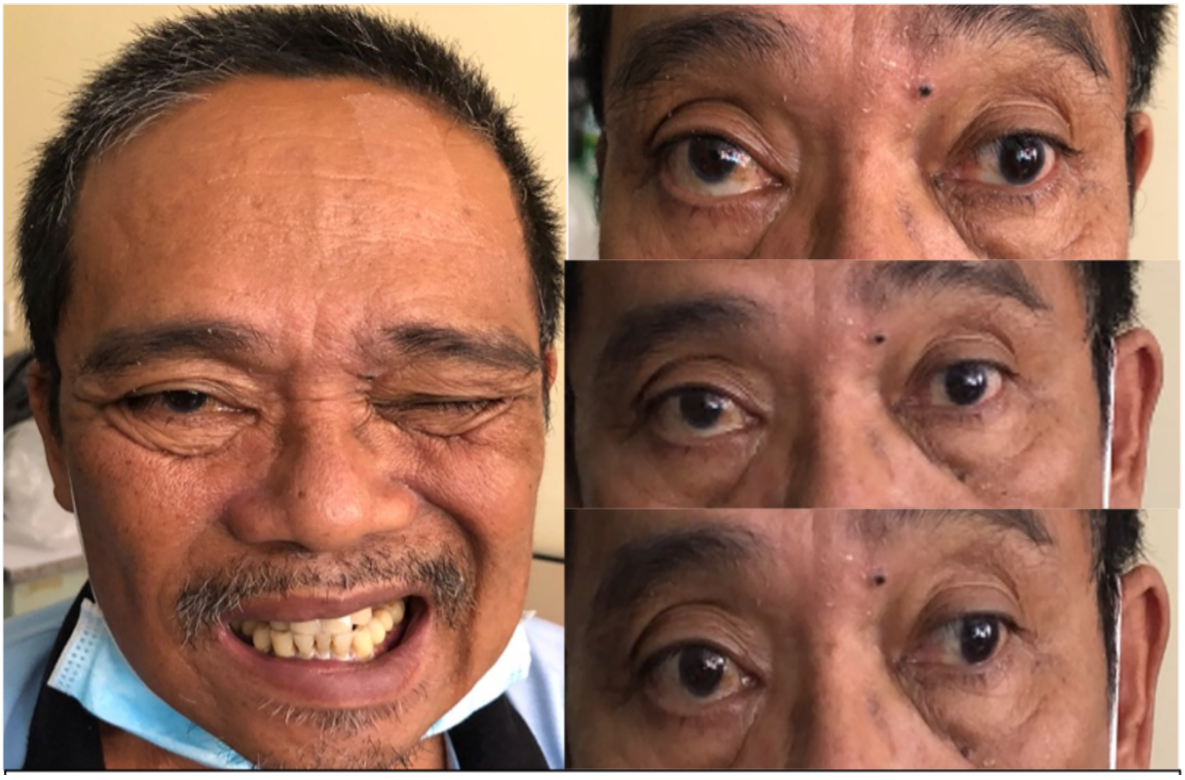
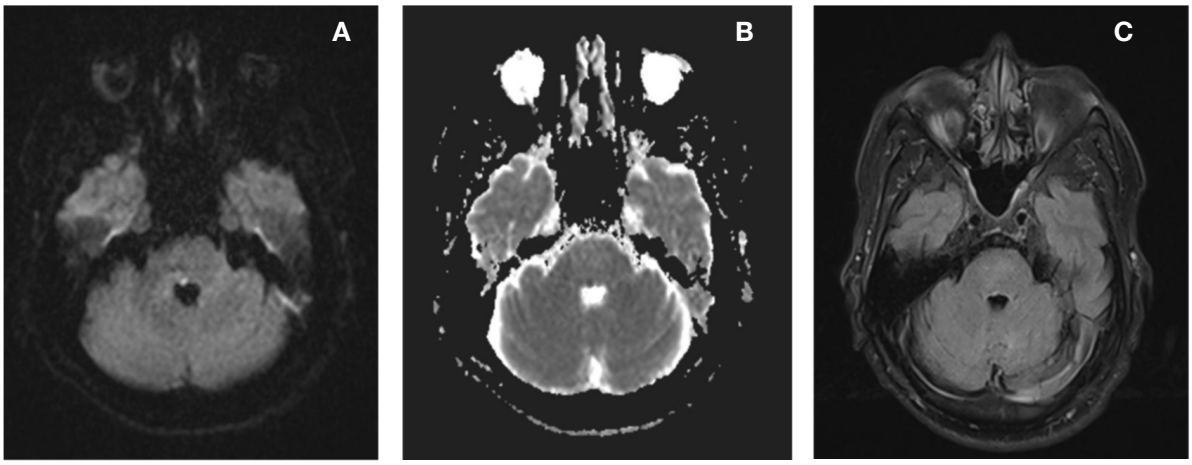


Figure 2a. Marked hyperintensity on diffusion weighted imaging (DWI) at the level of the facial colliculus, right dorsal pontine tegmentum.
Figure 2b. Signal reduction is evident on ADC at same area in the pons.
Figure 2c. Hyperintense signal on T2-FLAIR.



contralateral medial longitudinal fasciculus (MLF). A lesion at these connections would cause a horizontal gaze palsy of the ipsilateral eye and internuclear ophthalmoplegia (INO) or failure of adduction of the contralateral eye.[1,6,8] The only remaining eye movement in INO is abduction of the contralateral eye. These structures lie in close proximity to the nucleus and intraaxial fascicles of cranial nerve VII manifesting as facial weakness of the ipsilateral side to the lesion.[1,6,8] These structures are located in the dorsal pontine tegmentum which are vulnerable to a vascular event or demyelination, with an occlusion in the tip of the paramedian pontine artery, a branch of the basilar artery, as the most common etiology.[2,6] Space occupying lesions are reported although rare.[2, 8]

The most common causes of posterior circulation strokes are occlusion or embolism from large artery vertebrobasilar atherosclerosis or dissection, and cardioembolic events.[7]

Paramedian pontine infarcts are usually due to thrombosis of perforating arteries with outcomes usually based at the level of the pontine lesion. Poor outcomes are seen when the corticospinal tract is involved, leading to immobility.[9] In this case, the corticobulbar tract was involved but spared the corticospinal tract. Hypertension, diabetes, hyperlipidemia and smoking are associated risk factors for paramedian pontine infarcts.[7, 9]

CONCLUSION

Diagnosis of eight and a half syndrome is clinical, with radiographic findings acting as supplemental to the diagnosis. It is crucial for early detection of this kind of stroke syndrome, as its location is in the brainstem. Proper localization must be arrived at for sound diagnostics and treatment strategy.

REFERENCES

1. Duffy A, Shen PY, Verro P, Nidecker AE, Chow M. Eight-and-a-half syndrome. *Neurol Clin Pract*. 2014;4(6): 526–527. doi:10.1212/CPJ.0000000000000088
2. Sampath Kumar NS et al. Eight-and-a-Half Syndrome: A Rare Presentation of Pontine Infarction. *Journal of Stroke and Cerebrovascular Diseases , Volume 23 , Issue 8 , e389 - e391*. doi: 10.1016/j.jstrokecerebrovasdis.2014.01.002
3. P. Kakar, Z. Brown, S. Banerjee, Eight-and-a-half syndrome: an unusual presentation of brainstem infarction, *QJM: An International Journal of Medicine*, Volume 106, Issue 3, March 2013, Pages 273–276, https://doi.org/10.1093/qjmed/hcr187
4. Wall M,Wray SH. The one-and-a-half syndrome: a unilateral disorder of the pontine tegmentum: a study of 20 cases and review of the literature. *Neurology*. 1983;33:971-980
5. Eggenberger E. Eight-and-a-half syndrome: one-and-ahalf syndrome plus cranial nerve VII palsy. *J Neuroophthalmol*. 1998;18:114-116
6. Radzuan IS, Hooi O, et.al. Eight-and-a-Half Syndrome: A Rare Presentation of Gaze Palsy Following Cerebrovascular Event. *Sch J Med Case Rep*. 2017; 5(10): 606-609
7. Merwick A, Werring D. Posterior circulation ischaemic stroke. *BMJ*. 2014 May 19;348:g3175. doi: 10.1136/bmj.g3175
8. Mesina BVQ, Sosuan GMN, Reyes KB. Eight-and-a-half syndrome: a rare potentially life-threatening disease. *GMS Ophthalmol Cases*. 2018;8:Doc04. Published 2018 Mar 8. doi:10.3205/oc000086
9. Kataoka S, Hori A, Shirakawa T, Hirose G. Paramedian pontine infarction. Neurological/topographical correlation. *Stroke*. 1997 Apr;28(4):809-15