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Interesting Case of Opalski Syndrome a Variant of Lateral Medullary Syndrome.

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ABSTRACT

Lateral medullary (or Wallenberg's) syndrome is characterized by vertigo, diplopia, dysarthria, Horner`s syndrome, numbness (ipsilateral face and contralateral limb) and traditionally it is not associated with any limb weakness. Localization in this syndrome is easy because of characteristic presentation, exclusive blood supply and very small area of involvement. In our case patient had features of lateral medullary syndrome with ipsilateral hemiparesis .In Opalski syndrome hemiplegia is ipsilateral due to the extension of the infarct caudally to involve the corticospinal fibers after the pyramidal decussation. Opalski syndrome is a rare variant of Wallenberg's syndrome with ipsilateral hemiparesis. It should not be diagnosed in the absence of hyperreflexia or Babinski sign. Excessive alcohol intake may play a role in the etiopathogenesis of posterior circulation stroke.

Keywords: Hemiparesis, Lateral Medullary Syndrome, Opalski syndrome, Wallenberg's syndrome, Babinski-Nageotte syndrome , horners syndrome , nystagmus ,

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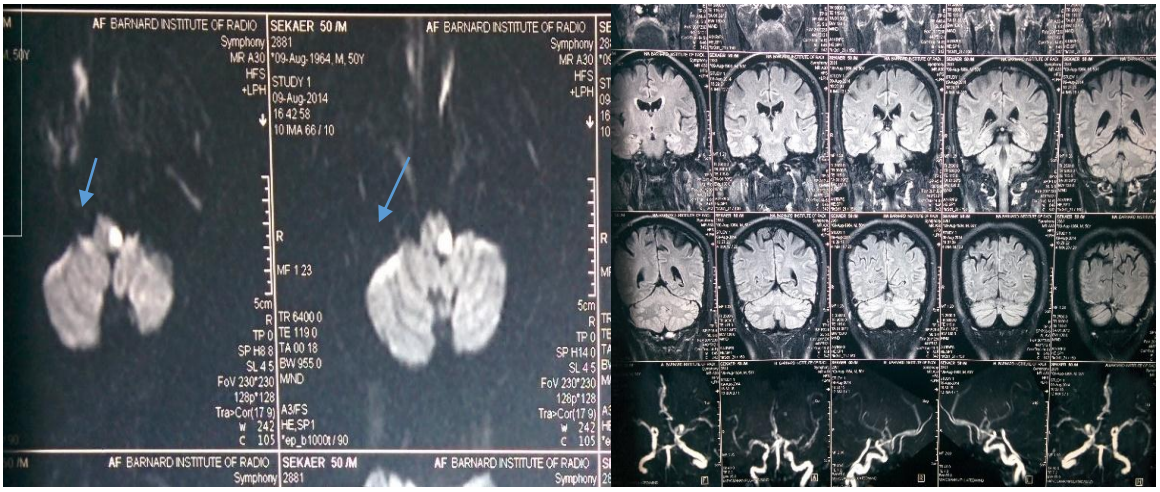
INTRODUCTION

Lateral medullary (or Wallenberg's) syndrome is characterized by vertigo, diplopia, dysarthria, Horner`s syndrome, numbness (ipsilateral face and contralateral limb) and traditionally it is not associated with any limb weakness. Localization in this syndrome is easy because of characteristic presentation, exclusive blood supply and very small area of involvement. However in Opalski syndrome [1] and Babinski-Nageotte syndrome [2], lateral medullary syndrome is associated with hemiplegia. In Opalski syndrome hemiplegia is ipsilateral due to the extension of the infarct caudally to involve the corticospinal fibers after the pyramidal decussation [1]. In Babinski-Nageotte syndrome [2] there is contralateral hemiparesis because pyramidal tract is affected before decussation.

We report a case of 50 yr old gentleman diabetic and hypertensive, presented to opd with complaints of giddiness three days, slight weakness of left upper and lower limb 3days. Numbness over left side of face and right upper limbs & lower limbs 3days.patient was apparently normal 3days before , he developed giddiness which is acute in onset associated with tinnitus , mild dysphagia to solids ,dysarthria which lasted for a day but vertigo was persistent in nature .patient gives history of mild weakness of left upper and lower limbs , unsteadiness while walking , decreased perception of sensation over right upper limbs and lower limbs , spillage of water while drinking more on left side , slight deviation of angle of mouth to right, numbness over left side of face . No history of cough, hoarseness of voice, neck pain, no evidence of headache or vomiting and no fever during the period of illness. No bowel and bladder disturbance, no h/o trauma, seizures. Diabetic for the past 5yrs on OHA's, hypertensive for the past 4yrs on tab.amlodipine 5mg once a day. No other significant past history. Family history not significant. Alcoholic and smoker for the past 20 yrs . Consumes approximately 360 ml of brandy per day thrice or more in week and a bundle of beedi per day. On examination moderately built and nourished pulse 84bpm , regular BP 130/80 mm hg left upper limb in sitting position 130/80 mm hg right upper limb in sitting position . Patient is not pale / icteric ,no clubbing no cyanosis no edema no lymphadenopathy no neuro-cutaneous markers ,HMF – conscious , oriented ,right handed , memory intact , dysarthria , cranial nerves - loss of pain and temperature over left half of the face , left side umn 7th cranial nerve palsy ,left horners syndrome (ptosis,miosis, decreased sweating) , nystagmus seen rotatory left side , gag reflux diminished on left side , uvula deviated to right side.

MRI brain with MRA →features suggestive of acute infarct in left lateral medulla



Spino motor system(reveled left hemiparesis) bulk was normal on both sides , tone appears normal on both sides , power of 4/5 in both upper and lower limbs on left side with normal power on right side , reflexes brisk on left side upper and lower limbs with normal reflexes on right side , plantar extensor on left side .No involuntary movements, ataxic gait.

Sensory loss over right half of body which included loss of pain and temperature sensation. No sensory loss on left half of body, vibration and joint position sense were intact on both sides. Cortical sensation intact on both sides .cerebellar signs are present on left side. Spine and cranium normal, no signs of meningeal

irritation, no trophic changes. Lungs were clear and heart sounds were normal .abdomen was soft, non-tender, no hepatomegaly no splenomegaly, bowel sounds were heard normally.

Laboratory investigations revealed normal complete blood counts, esr with in normal limits, normal renal function, normal serum electrolytes. Urine routine analysis was normal, Liver function test revealed mild elevation of GAMA GGT, SGOT, SGPT. USG abdomen revealed fatty liver. Blood sugars were within limits. Lipid profile was impaired.

ECG was normal, 2d echo normal study , doppler study of carotids and vertebral artery was normal.

This patient presented with all features of lateral medullary syndrome. In addition patient had ipsilateral hemiparesis (corticospinal tract involved) it is a variant of lateral medullary syndrome – opalski syndrome type 1 sensory variant.

He was reassured started on tab.aspirin 150mg once a day , tab.rosuvastatin 10mg hs, Tab.citicoline 500mg twice a day , tab.enalapril 2.5mg twice a day ,continued oral hypoglycemic drugs ,physiotherapy and diet management. Neurologist consultation was done. His general condition gradually improved and is in follow up with us.

DISCUSSION

Lateral Medullary Syndrome (LMS) was described by Wallenberg [3]. It is a relatively uncommon stroke with slight male sex preponderance [3,4]. The classical presentation consists of crossed sensory deficits, specifically loss of pain and temperature sensation affecting trunk and extremities contralateral to the infarct along with ipsilateral facial numbness. Other features of the syndrome include vertigo, nystagmus, hoarseness, dysphagia, ipsilateral cerebellar signs and Horner's syndrome [4].

In most cases the diagnosis of LMS is made on the basis of classical clinical features and some authors even recommend delaying the MRI scan when the presentation is classical [5]. However, a wide variation has been recognised in the clinical presentation [5,6] and studies have demonstrated that the classical presentation is not seen most of the time. Hence, efforts have been made to classify LMS on the basis of pattern of sensory involvement [7], anatomical localisation [8] and radiological pattern of involvement [3].

The wide variation in sensory involvement has long been recognised and Zhang et al., described five patterns of sensory impairment [7].

Type 1: Ipsilateral face and contralateral trunk and limbs
Type 2: Ipsilateral face and contralateral face, trunk and limbs
Type 3: Contralateral face and body
Type 4: Ipsilateral face and contralateral trunk and leg
Type 5: Contralateral face, arm and upper trunk

Our patient would come under classical or type 1 sensory involvement, as he presented with left sided face and right sided trunk and limb involvement. In clinico radiological correlation studies by Kim et al., only 26% of the patients presenting with the classical crossed sensory pattern or Zhang type 1 and 25% presented with Zhang type 2.This variation has been attributed to the different patterns of involvement of medulla classified horizontally as dorsal, ventral and lateral and vertically as rostral and caudal [4]. The more rostral lesions which tend to be more ventral presented with contralateral trigeminal nerve involvement while the more caudal and dorsolateral lesions tend to present classically as seen in our case [3].

The absence of the classical component of Wallenberg's syndrome – Horner's syndrome is also explained by this differential involvement. Kim et al., have demonstrated that Horner's syndrome is uncommon when the dorsal part of medulla is involved, as seen in our patient [4]. It was also observed that dorsolateral lesions tend to be more superficial and hence do not involve nucleus ambiguous, which is situated more deeply [3]. This explains the absence of dysphagia and signs of palatal paralysis in our patient. The spontaneous facial pain experienced by our patient is part of some of the original descriptions of Wallenberg syndrome. It has been described as a burning type of pain which usually comes with the onset of symptoms. It

has been attributed to involvement of phylogenetically older pain pathway which receives fibres from both the spinothalamic tract and the trigeminal nerve [8].

The other additional feature in our patient was the presence of ipsilateral hemiparesis. This variant of Wallenberg's syndrome was first described by Opalski in 1949[1]. Wallenberg's syndrome with contralateral hemiplegia is called as Babinski Nageotte syndrome [2]. Some authors question the existence of hemiparesis as they attribute the weakness to represent a spinocerebellar hypotonic syndrome rather than represent a pyramidal tract involvement[3]. But the presence of Babinski sign in our patient suggests pyramidal tract involvement. In fact Hermann et al., have suggested that the eponym Opalski syndrome should be reserved only for those cases with ipsilateral hemiparesis with Babinski positivity and contralateral numbness [10].

The cause of hemiparesis remains controversial. Opalski attributed it to caudal extension of the infarct to involve corticospinal fibres after the decussation. But recent studies suggest that involvement of medullary penetrating arteries which supply the pyramidal fibres post decussation may be responsible. These arteries are a branch of the vertebral artery, which has been identified as being most common artery to be involved in Wallenberg's syndrome [9].

Alcohol is not an established risk factor for stroke, specifically posterior circulation stroke. But, a study from China has suggested that incidence of posterior circulation stroke is twice as high in alcoholics when compared to general population [11]. More detailed studies are needed before we can establish the role of alcohol in posterior circulation stroke, but it is reasonable to say that it might have played a role in our patient.

CONCLUSION

LMS can have a diverse clinical presentation and the absence of the classical pattern should not defer one from diagnosing Wallenberg syndrome. Radiology helps in understanding how vascular lesion localisation can lead to changed presentation. Opalski syndrome is a rare variant of Wallenberg's syndrome with ipsilateral hemiparesis. It should not be diagnosed in the absence of hyperreflexia or Babinski sign. Excessive alcohol intake may play a role in the etiopathogenesis of posterior circulation stroke.

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