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A Rare Case of Oculopalatal Myoclonus

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ABSTRACT
We present a rare case of a 40 year old female who suffered a pontine haemorrhage due to uncontrolled hypertension and a year later, was found to have oculopalatal myoclonus. Oculopalatal myoclonus is characterized by rhythmic pendular vertical eye movements associated with synchronous contraction of the soft palate. It produces intractable oscilopsia and normally is due to brain stem haemorrhage. The site of the lesion usually is in the triangle of GuillianMollaret. We would like to report this case due to its rarity. Most of these patients do no complain of dysphagia or dysphonia and should be counselled about that fact that they can lead a normal life despite having anoculopalatal myoclonus.
Keywords-*oculopalatal myoclonus, guillianmollaret triangle.*

Introduction

Oculopalatal myoclonus is characterized by rhythmmic pendular vertical eye movements associated with synchronous contraction of the soft palate. [1] It produces intractable oscilopsia and is normally due to brain stem haemorrhage. [2] It does not manifest until several months or even years later with the longest recorded interval being 49 months. [3] We present a rare case of a patient who suffered a pontine haemorrhage due to uncontrolled hypertension and a year later she was found to have oculopalatal myoclonus.

Case

A 46 year old female, hypertensive on medication, non diabetic, attended our neurology outpatient department for follow up of her pontine haemorrhage with right hemiparesis which occurred a year back. Presently she complained of gradual progressive unsteadiness of gait, slurring of speech and in-coordination of limbs for the past 5 months. When she became ambulatory, while she was recovering from her pontineheamorrhage, she stated she had gait instability which was gradually progressive. At the same time she reported improvement of her muscle power of the paretic side. Gait instability was in the form of swaying to either side. No history of stiffness or abnormal involuntary movements affecting a limb. No diurnal variation of the unsteadiness. Slurring of speech as described by the patient caregiver was in the form of abnormal emphasis on particular syllables. No

history of inappropriate words or irrelevant talking. Patient gave a history of tremulousness in both upper limbs while reaching the object of interest. No tremulousness at rest. No slowness in initiating movements.

On examination, patient was alert, conscious co-operative. Blood pressure was 140/90 in left arm in supine position. Pulse of 86 per minute, regular in rhythm and all peripheral pulses where palpable.

Nervous system examination showed normal mental status. Spine and cranium was normal. Extra ocular movement examination showed spontaneous pendular eye movement in horizontal, vertical and oblique planes on primary gaze. The vertical movements were predominant on lateral gaze. There was adduction failure on right gaze but convergence was intact. Rest of the ocular movement range was within normal limits. Right nasolabial fold was less prominent and frontal creases were normal bilaterally. On examination of the palate, there were spontaneous involuntary soft palatal contractions bilaterally. On phonation, there was adequate elevation of the soft palate in spite of the continuous contractions and gag reflex was preserved. Other cranial nerves were within normal limits. Motor exam revealed slightly spastic weakness in the right upper and lower limbs. Power was 4-/5 on the right side. Jerks were asymmetric and slightly brisk on the right side. Right planter was extensor. Finger finger nose was impaired on both sides, dysdiado-kinesia present on both sides with left more than right. Romberg was negative. Patient had a right sided circumduction on walking and tandem was impaired.

Table 1: Investigations

Investigaton	Report
Haemoglobin	12gm/dl
White cell count	5300cmm (N71 L27 B00 E01)
ESR	40mm in 1 hr
Urea	26mg/dl
Creatinine	1.0mf/dl
Na	138
K	4.1
FBS	88mg/dl
PPBS	131mg/dl
ECG	LVH, LAHB
MRI Brain (1 year old)	Haemorrhage in upper pons region with peripheral edema
MRI brain (on admission)	Resolved heamorrhagic infarct in left Upper pons. No new lesion

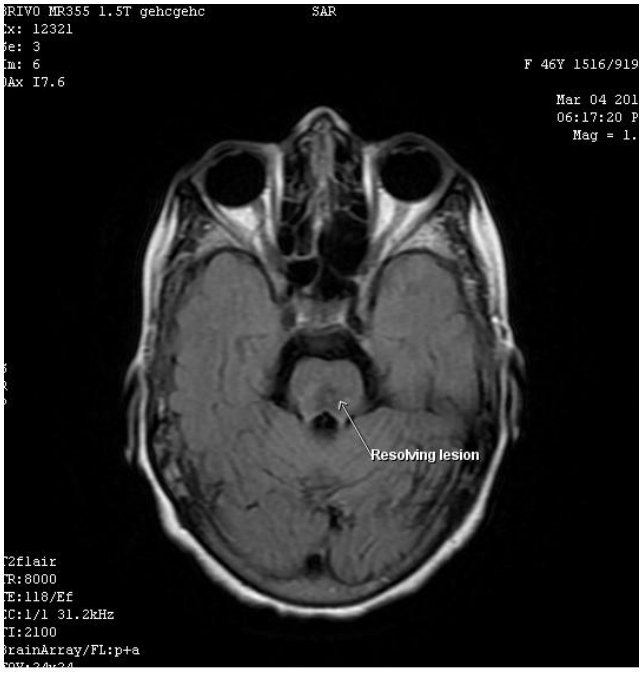

Figure 1: MRI brain showing resolving haemorrhagic infarct in left pons

Discussion

Palatal myoclonus (PM) is characterized by rhythmic involuntary jerky movements of the soft palate and also sometimes other muscles related to the pharynx. When associated with eye movements, it is called oculopalatal myoclonus (OPM). PM is exceptional among movement disorders because of its persistence during sleep and frequently lack of modulation by voluntary influences.

A syndrome similar to PM called essential palatal myoclonus (EPM) is a condition where there is no obvious finding on brain imaging. So PM are of two types, ordinary PM and EPM.

Ordinary PM is caused by a lesion in the triangle of Guillian Mollaret (1933). [4] Gullian and mollaret were French neurologists and perhaps this is why much of the literature is from French authors. This triangle is composed of the inferior olivary nucleus in the medulla, the central tegmental tract which connects the inferior olive to the red nucleus in the mid brain, the inferior cerebellar peduncle which connects the inferior olive to the cerebellum, the superior cerebellar penduncle which connects the red nucleus to the contralateral dentate nucleus of the cerebellum. [4]

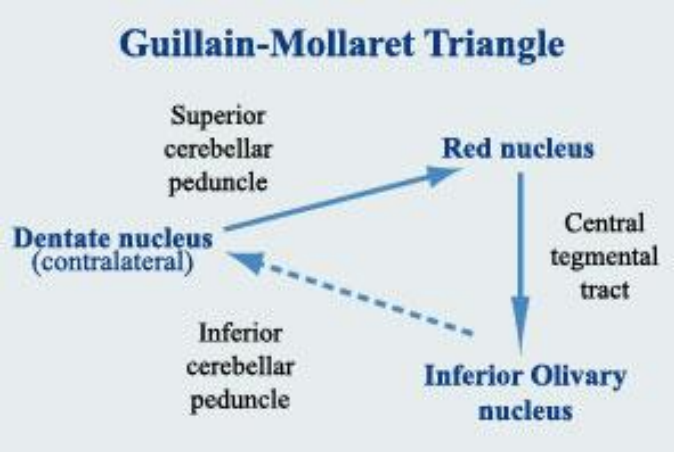

Figure 2: Guillain Mollaret Triangle

It is thought that the inferior olive enlarges and develops rhythmic discharges between 0.5-3 Hz, when it is denervated by ipsilateral brainstem disease or contralateral cerebellar disease and is responsible for the palatal myoclonus. [5] The inferior olive is provided with massive inhibitor (GABA) projections from the cerebellar nuclei. This differs from most other cerebellar output which is excitatory. Olivary neurons are extensively coupled by gap junctions which may account for their tendency to oscillate when inhibition is released. [5]

Cuases of lesion in this triangle are Stroke (40%), tumour (7%), trauma (8%), multiple sclerosis (8%), encephalitis (2%) and degenerative disease (2%). [6]

Generally no treatment works. In reported papers, about 20% of treatments have been successful, but some indicate that no treatment works.[7]

Ordinarily, the diagnosis is not difficult at all and mainly depends on the examiner looking at the palate and the eye. If one sees palatal oscillations, the options are very limted- OPM, EPM and functional syndrome. Distinction is made by associated findings. If eye moves with palate – OPM must be real and not stimulated. If MRI brain is abnormal with a lesion at Gullian Mollaret triangle, it should be a non essential variant of OPM as was our case.

We would like to report this case due to its rarity yet an easy diagnosis. Most of these patients do no complain of dysphagia or dyspnonia and should be counselled about that fact that they can lead a normal life despite having aoculopalatal myoclonus.

References

1. Leigh RJ, Zee DS. The neurology of eye movements. 3rd ed. New York: Oxford University Press, 1999.
2. Talks SJ, Elston JS. Oculopalatal myoclonus: eye movement studies, MRI findings and the difficulty of treatment. Eye 1997;11:19–24
3. Matsuo F, Ajax ET. Palatal myoclonus and denervation supersensitivity in the central nervous system. Ann Neurol 1979;5:72–8.
4. Guillain G, Mollaret P: Deuxcas de myocloniessynchrones et rythméesvélo-pharyngo-laryngo-oculo-diaphragmatiques: le problèmeanatomique et physio-pathologique de ce syndrome. Rev Neurol (Paris) 1931;2:545–566.
5. Mugnaini E, Nelson BJ. Corticotrophin-releasing factor (CRF) in the olivo-cerebellar system and the feline olivary hypertrophy. EBR Series 17, 187-197, 1989
6. Deuschel G, Mischke G, Schenck E, Schulte-monting J, Lucking CH. Symptomatic and essential rhythmic palatal myoclonus. Brain (1990), 1645-1672
7. Leigh, R. J., T. H. Burnstine, R. L. Ruff, et al. (1991). "Effect of anticholinergic agents upon acquired nystagmus: a double-blind study of trihexyphenidyl and tridihexethyl chloride." Neurology41(11): 1737-41.