


One and a half syndrome: A rare phenomenon in pontine infarction

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Introduction

One and a half syndrome is a rare ophthalmological phenomenon associated with pontine lesions. It could manifest either alone or in combination with other additional neurological symptoms. Isolated one and half syndrome is extremely rare. This case report describes about a patient presented with isolated one and half syndrome leading to subsequent finding of pontine infarction.

One-and-a-half syndrome (OHS) is caused by a lesion of unilateral tegmentum of pons, causing damage to the para pontine reticular formation (PPRF) and medial longitudinal fasciculus (MLF) (1). It is characterised by the combination of unilateral horizontal gaze paralysis and internuclear ophthalmoplegia (INO). The most common cause of OHS is vascular brainstem damage, mainly brain stem lacunar infarction. Demyelinating diseases and infections are also well recognised causes. Only a very small number of patients develop isolated OHS while the lesions are smaller and localised (1, 2). It is important to recognise OHS as early as possible which will help to localise the lesion and start proper treatment tailored to each patient.

Case presentation

A 72-year-old man, known patient with hypertension, ischaemic heart disease, bronchial asthma and diabetes mellitus was admitted to Teaching Hospital Jaffna (THJ), Sri Lanka with complaints of lethargy, restlessness, giddiness, headache, and visual disturbances for 2 days duration. He did not have recent onset of facial or

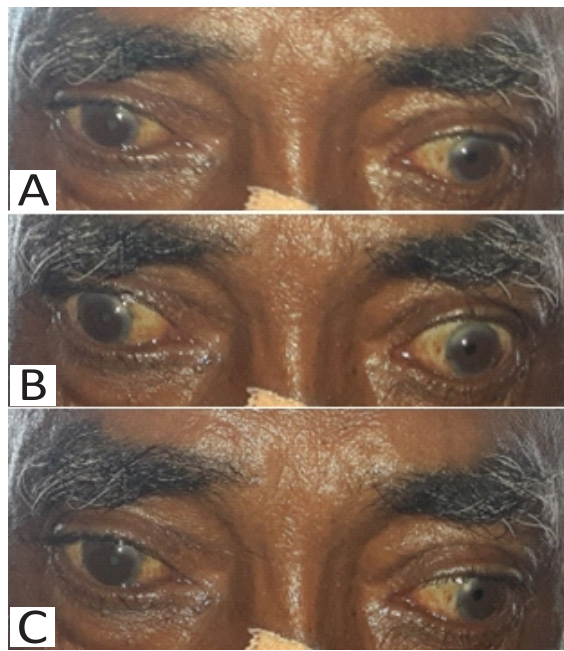
limb weakness. He denied any history of preceding fever or head trauma. He did not have constitutional symptoms such as loss of appetite or loss of weight.

On examination, he was found to be drowsy and had complete horizontal gaze palsy of right eye. Left eye abduction was preserved with nystagmus while adduction was lost. Vertical eye movements were preserved (Figure 1). Clinical diagnosis of OHS was confirmed by the subsequent Non contrast Computer Tomography (NCCT) of brain which revealed right side acute pontine infarction (Figure 2). Two-dimensional echocardiogram (2D Echo) ruled out intracardiac source of thromboembolism. His blood investigations were normal.

He was treated with Aspirin 75 mg daily, Clopidogrel 75 mg daily and Atorvastatin 40 mg daily all given orally and was discharged with same medications and clinic follow up was arranged at THJ.

Discussion

OHS consists of a gaze palsy in which one eye is completely unable to move horizontally, while on the other side adduction is absent and abduction is preserved (2). The only horizontal movement that remains is abduction of the contralateral eye, usually accompanied by nystagmus (3). Convergence is spared. When looking straight, the contralateral eye is slightly abducted which is called as paralytic pontine exotropia (Figure 1A). OHS is due to the lesion in the PPRF and/or abducens nucleus and MLF on the same side pons (4).



- A. On looking straight (Neutral gaze), the contralateral eye (Left) is slightly abducted (Paralytic pontine exotropia)
 B. On looking to right side, abduction of right eye and adduction of left eye, both are affected
 C. On looking to left side while the abduction of left eye is intact, adduction of right eye is affected

Figure 1: Position of eye on different instructions



Figure 2: NCCT brain shows focal infarction in right side pons (Yellow arrow)

The usual clinical features are diplopia, blurred vision and oscillopsia (2). Depending on the extent of the lesion, other neurological findings may be noted. Horner Syndrome, weakness or spasticity, sensory deficits, abnormally brisk or asymmetric reflexes, extensor plantar responses, and in coordination are other documented findings. If the facial motor nucleus is also affected, a phenomenon called “eight-and-a-half syndrome” appear in which there will be ipsilateral upper motor neuron type of facial nerve palsy in addition to the OHS. Other associations with OHS include oculopalatal myoclonus and Cheiro-Oral Syndrome (3).

Infarction of pons mainly due to occlusion of paramedian perforating branches of the basilar artery, demyelinating diseases such as multiple sclerosis (MS) and acute demyelinating encephalomyelitis (ADEM), haemorrhage, vascular malformations such as brainstem cavernous haemangioma and aneurysms, tumours (either primary or metastasis), hindbrain anomalies like Chiari malformation, and infectious lesions including neurocysticercosis, tuberculosis and brainstem encephalitis are common causes implicated in the development of OHS. Pontine infarction is the most common ethology in elderly population, while tumours and MS are common aetiology among younger patients (2, 3).

PPRF is located near the ipsilateral abducens nucleus. PPRF gives input to the ipsilateral abducens nucleus. The abducens nerve, which controls the ipsilateral lateral rectus muscle, and MLF, which give input to the contralateral oculomotor sub nucleus thus participating in contralateral eyeball adduction, originate from the ipsilateral abducens nucleus. So, horizontal conjugate movement, comprising ipsilateral abduction and contralateral adduction, is mediated by ipsilateral abducens nucleus which in turn is controlled by ipsilateral PPRF. If the PPRF or abducens nucleus is damaged, the eyes could not gaze at the lesion side, presenting the horizontal gaze palsy to the lesion side (1). If the abducens nucleus is spared, the gaze palsy is apparent on voluntary horizontal gaze, but is normal with involuntary oculocephalic manoeuvres, as PPRF participates in voluntary eye movement, but in the later it does not (4).

MLF receives the nerve impulses from visual area of the frontal and parietal lobes and participates in horizontal conjugate eye movements. Damage to the ipsilateral MLF results in ipsilateral adductor muscle paralysis and contralateral horizontal nystagmus, called as ipsilateral INO, when staring at the opposite side. If the damage does not involve the midbrain, the convergence reflex is integrated (1).

Hering's law of equal innervation, which states that any attempt to increase the innervation to a weak muscle in one eye must be accompanied by a commensurate increase in innervation to the yoke muscle in the other eye, under which increased innervation to the underacting adducting muscle would result in an enhanced stimulus to the contralateral abducting muscle is believed as the possible mechanism for the dissociated contralateral horizontal gaze evoked nystagmus in the abducting eye (5).

Magnetic resonance imaging (MRI) with diffusion weighted imaging are helpful in finding the aetiology of OHS including small ischaemic brainstem damage even in the acute stage. The lesions are usually located in the dorsomedial pontine tegmentum, contacting the floor of the fourth ventricle (2).

Patients with OHS caused by cerebrovascular disorders and multiple sclerosis have a good outcome. Majority of the patient will recover without major neurological sequelae (2).

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