MMJ

Malta Medical Journal, 35 (1): Pages (2023)

http://mmsjournals.org/index.php/mmj

CASE REPORT

One-and-a-half syndrome: Its presentation, causes and neuroanatomy

Matthew Azzopardi, David Agius

Background

One-and-a-half syndrome involves a combination of an ipsilateral horizontal gaze palsy and an ipsilateral internuclear ophthalmoplegia. This condition is easily missed due to its presentation, but can be the first sign of serious disease. We aim to increase awareness of this syndrome's presentation and give an insight into the intricate neuroanatomical connections that are affected in it.

Case Presentation

We present a case of a 39-year-old previously healthy female who presented with a one-week history of diplopia and non-vertiginous dizziness. On examination, a left horizontal gaze palsy with deficits in left abduction and right adduction was noted, accompanied by left adduction weakness and right horizontal disconjugate jerk nystagmus in abduction. A diagnosis of OAHS was made, and she was admitted for further tests. An MRI of her brain revealed multiple hyperintensities throughout, along with an enhancing lesion in keeping with active disease. A diagnosis of Multiple Sclerosis was made and she was given a five-day course of methylprednisolone, with which her vision, and ultimately her gait, improved. She was discharged with outpatient follow-up, to further discuss treatment options for her new diagnosis.

Conclusion

Diplopia and vertigo are symptoms that should prompt careful clinical examination with proper attention to ocular motility testing, and subsequent referral to neurology if required. Unnecessary delays in diagnosis and management could ultimately be detrimental to the patient, and being aware of uncommon presentations would go a long way in enhancing patient safety.

Matthew Azzopardi* MD (Melit.)
Royal London Hospital
Barts Health NHS Trust
London, UK
matthew.azzopardi.96@gmail.com

David Agius MD(Melit.) MRCS FEBO FRCOphth
Ophthalmologist and Lead Researcher
Faculty of Medicine and Surgery
Department of Surgery
Ophthalmology Outpatients
Mater Dei Hospital
Msida, Malta

**Corresponding Author*

Introduction

One-and-a-half syndrome (OAHS), first described in 1967,[1] presents with a combination of an ipsilateral horizontal gaze palsy (the 'one') and an ipsilateral internuclear ophthalmoplegia (INO) (the 'half').

Here we present a case of this syndrome which presented to Ophthalmic Casualty in Malta, in order to increase awareness of this syndrome's presentation and give an insight into the intricate neuroanatomical connections that are affected in this syndrome.

Case

A 39-year-old previously healthy female presented to ophthalmic casualty with a one-week history of diplopia and non-vertiginous dizziness. These were interfering with her daily life due to multiple falls, as well as with her job as a lifeguard due to a decreased ability to differentiate between different areas of the beach. She denied having other symptomatology such as headaches or nausea. She was prescribed prochlorperazine by her general practitioner, but her symptoms only got worse and she decided to go to the Emergency Department. Her past medical history included occasional sinusitis and myopia, for which she uses glasses. Her only past surgery was a lower segment caesarean section. She denied any smoking, drug use or excess alcohol intake, and was on no regular treatment. Her family history was unremarkable, and she was only allergic to seafood.

On examination, her aided visual acuity was 6/18 in each eye, which improved to 6/12 in either eye using pinhole. Pupils were equal and reactive to light, and no relative afferent pupillary defect was present. On attempted left gaze, there was a left horizontal gaze palsy (ie deficits in left abduction and right adduction) whilst on attempted right gaze, there was a left adduction weakness and right horizontal disconjugate jerk nystagmus in abduction. Convergence of the eyes was intact, and no other significant findings were noted on examination.

In view of her left lateral gaze palsy and left sided internuclear ophthalmoplegia, a diagnosis of OAHS was made. The on-call neurology team assessed her and admitted her for further investigations, to delineate the underlying cause.

Her blood investigations were all within normal limits, and a CT scan of the brain demonstrated no acute intracranial pathology. However, an MRI scan of her brain revealed a midline symmetrical hyperintensity underneath the floor of the fourth ventricle, along with multiple hyperintensities in both the cerebrum and cerebellum in a periventricular and subcortical distribution. An enhancing lesion in the left periventricular white matter was also noted, in keeping with an active plaque. These lesions were highly suggestive of demyelinating plaques, and a diagnosis of Multiple Sclerosis was made. She was given a five-day course of methylprednisolone, with which her vision improved and she started mobilizing independently. Alternating eye patching was also done to help with her vision. Once improved she was discharged with outpatient follow-up, to further discuss treatment options for her new diagnosis of multiple sclerosis.

Discussion

OAHS is a syndrome which manifests only in horizontal gaze. Three main structures are involved in horizontal gaze, namely the paramedian pontine reticular formation (PPRF), the internuclear fibres of the medial longitudinal fasciculus (MLF) and the abducens nuclei in the lower pons. Upon initiating horizontal gaze, an excitatory stimulus is sent from the PPRF to the ipsilateral abducens nucleus.[2] In turn, this trigger both the motor fibres of the

ipsilateral lateral rectus and also the contralateral medial rectus via the interneuronal fibres of the contralateral MLF, which ascend and terminate in the oculomotor nuclear complex.[3]

Since OAHS is purely a combination of ipsilateral horizontal gaze palsy and ipsilateral INO, a combination of lesions giving rise to these two components would need to be present. Firstly, with regards to the horizontal gaze palsy component, there are four theoretically possible lesion locations:

1. Damage to the ipsilateral abducens nucleus and ipsilateral PPRF
2. Damage to the ipsilateral PPRF only
3. Damage to the ipsilateral abducens nucleus only
4. Two separate lesions damaging the ipsilateral abducens nerve root fibre and the contralateral MLF[3]

Furthermore, the pattern of horizontal gaze palsy would also depend on the exact location of the lesion. In situations where ipsilateral PPRF damage is the culprit, both saccadic and pursuit eye movements are lost in addition to the horizontal gaze palsy. However, if the PPRF lesion is rostral to the abducens nucleus, vestibular reflexic horizontal eye movements are preserved, contrasting with PPRF lesions located at the level of the abducens nucleus, in which these reflexes are also lost. All of these voluntary and reflexic eye movements are also lost if the horizontal gaze palsy is due to ipsilateral abducens nucleus damage.[3-4]

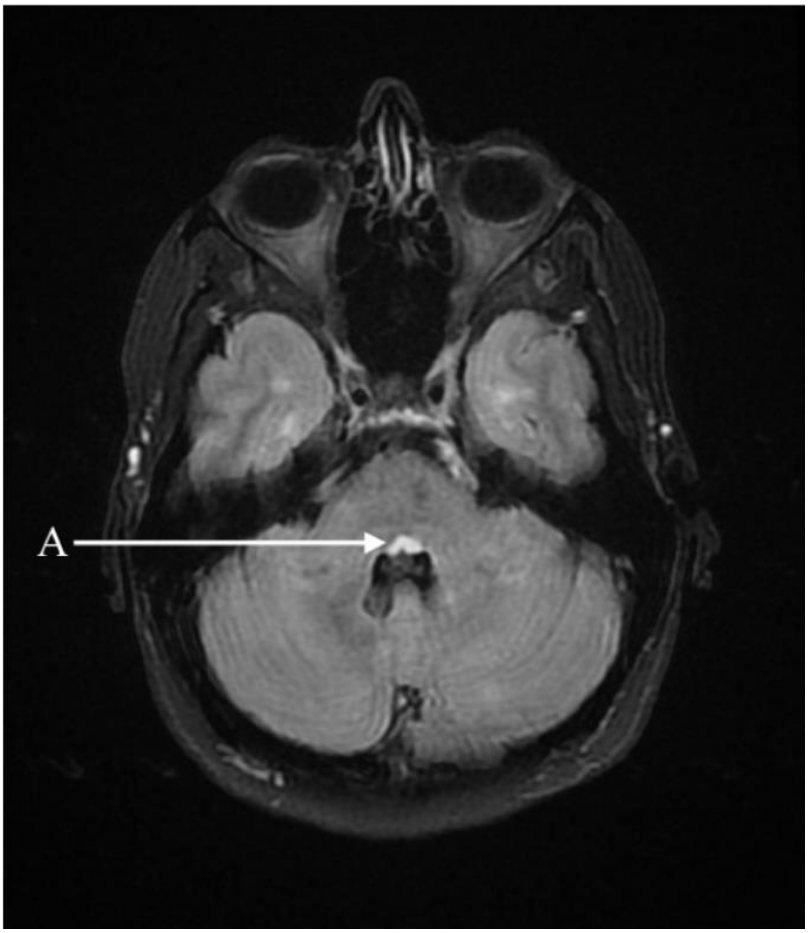
Secondly, the ipsilateral INO component of OAHS occurs due to lesions involving the ipsilateral MLF, in combination with those causing the horizontal gaze palsy. This occurs because of the integral role the internuclear fibres of the MLF play in conjugate horizontal eye movements. Because of this damage, the ipsilateral eye would be unable to adduct, and on abducting the contralateral eye a horizontal jerk nystagmus is observed, giving rise to INO.[3]

In fact, amongst the lesions found on MRI in our patient, a midline symmetrical hyperintensity underneath the floor of the fourth ventricle was noted, corresponding to the location of the colliculus facialis and the abducens nucleus (Figure 1).[5] This lesion was probably the cause of the horizontal gaze palsy component of her OAHS.

The commonest visual symptoms reported in OAHS include diplopia, blurred vision and oscillopsia.[3] However, it often presents with non-visual symptoms which would arise either as a consequence of the underlying pathology or of the visual disturbance itself, such as nausea, vertigo, and unsteadiness.[3] This highlights the importance of not dismissing non-specific symptoms and signs without a proper, thorough examination.

Many different disease pathologies can give rise to the pontine lesions behind OAHS. In our patient, OAHS was the first presenting feature of multiple sclerosis (MS). When one considers ocular presenting features of MS, optic neuritis immediately comes to mind. However, this is not the only possible ocular presenting feature, and in fact there are a number of other cases reported in which MS presented with OAHS.[3] Most OAHS cases are due to vascular causes, such as ischaemic or haemorrhagic brainstem infarction, basilar artery aneurysms and arteriovenous malformations. Other causes such as brainstem malignancies, infiltrative lesions, and infections have also been reported.[6]

Figure 1: Transverse section of the Magnetic Resonance Imaging scan of our patient's brain showing the midline symmetrical hyperintensity underneath the floor of the fourth ventricle corresponding with the abducens nucleus within the colliculus facialis (A)



Treatment of this syndrome is usually aimed at the underlying cause, as in our case. However symptomatic treatment modalities for diplopia, oscillopsia, blurred vision and other symptoms can be employed, such as eye patching or prism use. Botulinum toxin injections in the lateral rectus muscle and surgical extraocular muscle recession have also been used with good effect.[7-8]

Ever since this syndrome was first reported, similar syndromes have been described. For example, eight-and-a-half syndrome refers to one-and-a-half syndrome with ipsilateral facial nerve palsy,[9] whilst sixteen-and-a-half syndrome refers to eight-and-a-half syndrome with ipsilateral hearing loss (VIII cranial nerve).[10]

Conclusion

One-and-a-half syndrome is one of the lesser known ocular manifestations of common diseases such as multiple sclerosis and stroke. Diplopia and vertigo are symptoms that should prompt careful clinical examination including attention to ocular motility testing, with subsequent referral to neurology if required. In many of the pathologies that cause this syndrome, unnecessary delays in diagnosis and management could be detrimental to the patient. Proper examination and early involvement of the relevant specialities could go a long way in helping ensure the best possible outcome for our patients.

References

1. Fisher, C.M., Some neuro-ophthalmological observations. J Neurol Neurosurg Psychiatry, 1967. 30(5): p. 383-92.
2. Cohen, B. and A. Komatsuzaki, Eye movements induced by stimulation of the pontine reticular formation: evidence for integration in oculomotor pathways. Exp Neurol, 1972. 36(1): p. 101-17.
3. Wall, M. and S.H. Wray, 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(8): p. 971-80.
4. Azevedo Júnior, D., Pontine tegmentum hematoma: a case report with the "one-and-a-half" syndrome without pyramidal tract deficit. Arq Neuropsiquiatr, 1995. 53(3-a): p. 475-80.
5. Ettl, A. and E. Salomonowitz, Visualization of the oculomotor cranial nerves by magnetic resonance imaging. Strabismus, 2004. 12(2): p. 85-96.
6. Xue, F., et al., One-and-a-half syndrome with its spectrum disorders. Quant Imaging Med Surg, 2017. 7(6): p. 691-697.
7. Buckley, S.A. and J.S. Elston, Surgical treatment of supranuclear and internuclear ocular motility disorders. Eye (Lond), 1997. 11 (Pt 3): p. 377-80.
8. Kipioti, A. and R.H. Taylor, Botulinum toxin treatment of "one and a half syndrome". Br J Ophthalmol, 2003. 87(7): p. 918-9.
9. Eggenberger, E., Eight-and-a-half syndrome: one-and-a-half syndrome plus cranial nerve VII palsy. J Neuroophthalmol, 1998. 18(2): p. 114-6.
10. Cummins, G., et al., "Sixteen and a half": a novel pontine neuro-ophthalmological syndrome. J Neurol, 2011. 258(7): p. 1347-8.