

Isolated trochlear palsy as the only presentation of midbrain infarction: a case report

Journal of International Medical Research
49(4) 1–6

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DOI: 10.1177/03000605211008292

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Abstract

Trochlear palsy often results from traumatic, congenital and microvascular disorders. An intra-axial lesion as a cause of trochlear palsy is uncommon. Moreover, it usually accompanies other neurological deficits. Isolated trochlear palsy as the only presentation of brainstem stroke is unexpected. This current case report describes a 74-year-old male that presented with trochlear palsy without other neurological signs. Brain magnetic resonance imaging (MRI) revealed an acute midbrain infarction. The case report also reviews recent literature and provides a stepwise algorithm for clinicians to approach patients with trochlear palsy. Despite its rarity, clinicians are advised to consider ischaemic stroke as a cause of trochlear palsy even without other neurological deficits. Early MRI should be performed for prompt and proper management.

Keywords

Isolated trochlear palsy, midbrain infarction, intra-axial lesion, stepwise algorithm, magnetic resonance imaging

Date received: 25 November 2020; accepted: 16 March 2021

Introduction

Ocular motor palsy is the most common cause of diplopia.¹ Among the group of acquired ocular motor nerve palsies, abducens nerve palsy is the most prevalent (40%) followed by oculomotor nerve palsy (27%), trochlear nerve palsy (23%) and multiple ocular motor nerve palsies (10%).²

Since the superior oblique muscle that is innervated by the trochlear nerve mediates intorsion and depression of the eye, the

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diplopia is usually aggravated by the downward and contralesional gaze in patients with trochlear palsy. The aetiologies of trochlear palsy include congenital defect, trauma and microvascular neuropathy.¹ Brainstem stroke is an unexpected cause of trochlear palsy and usually accompanies other neurological deficits.³ Isolated trochlear palsy as a presentation of brainstem stroke is rare.³ This current case report describes a patient with midbrain infarction presented as isolated trochlear nerve palsy.

Case report

In February 2012, a 74-year-old Asian male with a past medical history of hypertension, type 2 diabetes mellitus, and bladder tumour post-operation, without tobacco or alcohol use, presented to the Emergency Department, Kaohsiung Medical University Hospital, Kaohsiung due to an acute onset of binocular vertical diplopia for 1 week. The diplopia was aggravated during leftward and downward gazing, making it hard for him to walk down the stairs. The course of his condition was stable without diurnal change. He did not report fever, headache, orbital pain or a history of trauma. Neurological examination revealed hypertropia of the right eye in the primary position and a positive Bielschowsky head tilt test: worsening of the hypertropia with a right head tilt and improvement with a left head tilt. These findings were compatible with right trochlear palsy. Other neurological signs were negative. Laboratory evaluation showed euthyroidism and poor blood sugar control (glycosylated haemoglobin: 9.6%).

Diffusion weighted imaging (DWI) revealed acute ischaemic stroke over the left aspect of the midbrain, close to the median plane, the location of the trochlear nucleus and intra-axial trochlear nerve (Figures 1a and 1b), which perfectly correlated with the clinical manifestations.

Figure 1c presents a schematic diagram of the relative location between the lesion site and the trochlear nucleus. No obvious lesions were identified in the other magnetic resonance imaging (MRI) scans (Figures 1d, 1e and 1f). Magnetic resonance angiography (MRA) reported atherosclerosis of the bilateral internal carotid arteries and vertebralbasilar arteries (Figure 1g). Carotid artery ultrasonography showed bilateral common carotid artery plaques without turbulent flow. No significant arrhythmia was found by electrocardiogram. The lipid profiles were within normal limits. The patient was prescribed 100 mg aspirin orally once daily for the prevention of secondary stroke. In addition, the antidiabetic agents were adjusted to include 750 mg metformin orally twice a day and 3 mg glimepiride orally twice a day for blood glucose control. The diplopia improved gradually 3 days later.

Written informed consent was obtained from the patient for publication of this case report. Steps have been taken to anonymize his details.

Discussion

In patients with binocular vertical diplopia, trochlear palsy is one of the most common causes.¹ In addition, skew deviation, ocular myasthenia gravis and thyroid ophthalmopathy may also result in vertical diplopia.¹ With regard to the diagnosis of trochlear palsy, a more sensitive two-step Parks–Bielschowsky test was proposed to replace the original three-step test.⁴ The two-step test includes hypertropia of the affected eye and the Bielschowsky head tilt test, with worsening of the hypertropia in the ipsilateral head tilt and improvement in the contralateral head tilt; that is, eliminating step 2 from the three-step test, which increased the sensitivity from 70% to 84% while not significantly decreasing the specificity of the test.⁴ The Bielschowsky head tilt

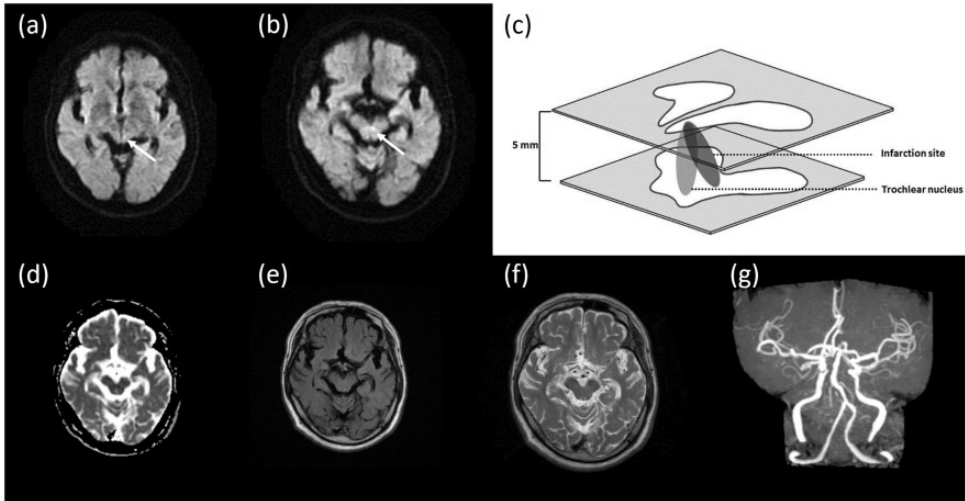


Figure 1. Magnetic resonance imaging (MRI) scans and an illustrated schematic diagram of a 74-year-old Asian male that presented with acute onset of binocular vertical diplopia: (a) diffusion weighted imaging (DWI) showed high signal intensity over the medial posterior portion of the left midbrain (white arrow); (b) DWI cut at the caudal side of the previous image showed high signal intensity over the medial anterior portion of the left midbrain, close to the interpeduncular fossa (white arrow); (c) schematic diagram of the relative location between the lesion site and the trochlear nucleus; the ischaemic lesion involved the trochlear nucleus with no obvious lesions identified in the other sequences, including in apparent diffusion coefficient (d), fluid attenuated inversion recovery (e) and a T2 weighted image (f); (g) brain magnetic resonance angiography showed atherosclerosis of the bilateral internal carotid arteries and vertebrobasilar arteries.

test could help physicians to discriminate trochlear palsy as the cause of vertical diplopia from skew deviation, which is a vertical strabismus caused by a supranuclear lesion in the posterior fossa.⁵ In addition, myasthenic serology and thyroid status tests can help to rule out myasthenia gravis and hyperthyroidism. Decompensation of congenital palsy, head trauma and microvascular neuropathy are the most common causes of isolated trochlear palsy in adults.^{1,6} Congenital palsy may not present in childhood if adaptations and subsequent decompensation occur later in life.⁶ Decompensation of trochlear palsy is difficult to identify, but it has been proposed that hypertropia greater in the upgaze than the downgaze or equal in the upgaze and downgaze is a characteristic of patients with decompensated congenital

palsies, which distinguish a congenital cause from the other aetiologies.⁷

Microvascular neuropathy associated with diabetes mellitus, hypertension or vasculitis is the most common cause of non-traumatic acquired trochlear palsy.⁸ Diabetic ophthalmoplegia was considered in this current patient who denied a history of trauma and had poor blood glucose control. However, the most frequently affected nerves in diabetes mellitus are the abducens and oculomotor nerves; the trochlear nerve is the least often involved.⁹ In addition, vascular events of the central nervous system share similar vasculopathological risk factors with microvascular damage; therefore, the possibility of a cerebrovascular accident should be taken into consideration. A previous prospective study compared the presumed microvascular cause and other rare

causes of isolated ocular motor nerve palsy.¹⁰ Even though presumed microvascular ischaemia accounts for the majority, the authors emphasized the importance of using MRI to identify other rare causes, such as brainstem infarction, intracranial neoplasm, aneurysm, inflammation and infection.¹⁰

An intra-axial lesion is an uncommon aetiology in patients with trochlear palsy without other neurological deficits.³ Because the trochlear nucleus and fascicles are surrounded by the ascending trigeminothalamic tract, spinothalamic tract, medial

longitudinal fasciculus, descending sympathetic tract and decussating fibres of the superior cerebellar peduncle, if an intra-axial lesion exists, it is usually accompanied with internuclear ophthalmoplegia, Horner's syndrome, upbeat nystagmus, sensory disturbances and ataxia.¹¹ Only a few cases of isolated trochlear palsy caused by intra-axial lesions have been reported.¹¹⁻¹⁴

Despite its rarity, these cases highlight the importance of neuroimaging. For patients with isolated trochlear palsy, MRI is superior to computed tomography because it is more sensitive in the detection of

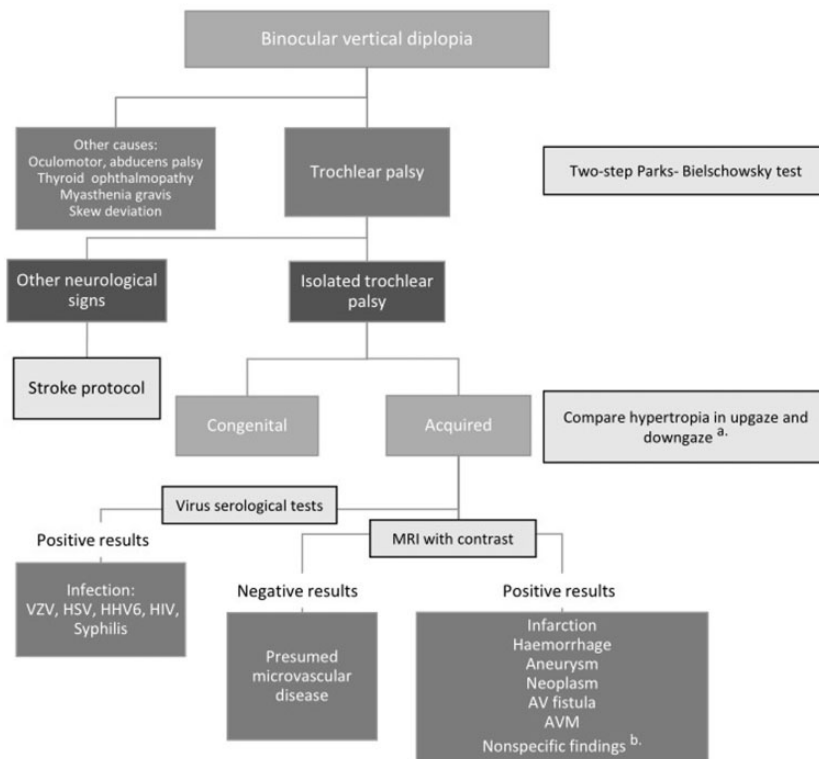


Figure 2. Stepwise algorithm to identify the aetiologies of trochlear palsy in adults: (a) if hypertropia is greater in the upward gaze than in the downward gaze or equal in the upward gaze and downward gaze, congenital trochlear palsy could be diagnosed; (b) if the magnetic resonance imaging (MRI) is abnormal, but the findings are nonspecific, then an autoimmune disease (e.g. neurosarcoidosis, Sjögren syndrome, systemic lupus erythematosus) should be taken into consideration. VZV, varicella zoster virus; HSV, herpes simplex virus; HHV, human herpesvirus; HIV, human immunodeficiency virus; AV, arteriovenous; AVM, arteriovenous malformation.

intracranial lesions in the posterior fossa. Early neuroimaging is essential to discover an underlying aetiology because subsequent management may be different and sometimes emergent. In this current case, a mid-brain infarction was found by MRI and antiplatelet therapy was given immediately for the prevention of recurrent stroke. Nevertheless, about 17% of patients with ischaemic stroke have lesions that are invisible on the DWI scan within 24 hours, especially when the lesions are small and located at the brainstem.¹⁵ In order to improve sensitivity, it is recommended to combine conventional axial DWI and sagittal DWI, which provide better coverage of the whole length of the brainstem.^{16,17} In the case of an initial negative finding, sagittal DWI may be added to improve the sensitivity for small-sized brainstem infarction while repetitive brain MRI could also be considered after 24 hours of onset.

As a consequence of their experience, the current authors provide a stepwise algorithm for clinicians to approach patients with trochlear palsy to identify the possible aetiologies (Figure 2). Firstly, the two-step Parks–Bielschowsky test can help to make the diagnosis of trochlear palsy.⁵ If non-traumatic isolated trochlear palsy is noted, it is essential to discriminate congenital causes from acquired causes by comparing hypertropia in the upgaze and downgaze.⁷ Neuroimaging is not absolutely necessary for congenital trochlear palsy. In addition, a viral serological test should be done to rule out infectious causes. Ultimately, MRI with contrast enhancement should be performed. If there is no finding on the MRI, plus the patient has at least one vascular risk factor and spontaneous resolution, microvascular disease could be diagnosed.² In rare cases, MRI may show an intra-axial lesion, including infarction, haemorrhage, aneurysm, neoplasm, arteriovenous fistula or arteriovenous malformation.^{11–14} If trochlear palsy cannot be

attributed to any cause after following the flowchart, or nonspecific findings were identified in the MRI, autoimmune disease (e.g. neurosarcoidosis, Sjögren syndrome, systemic lupus erythematosus) should be taken into consideration.^{18,19} In conclusion, the findings of this current case report suggest that clinicians should consider brainstem infarction as a cause of trochlear palsy even without other neurological deficits; and early MRI should be performed because the following management may be heterogeneous.

Declaration of conflicting interest

The authors declare that there are no conflicts of interest.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

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