Vertical gaze palsy in Parkinsonism: An alternative diagnosis to progressive supranuclear palsy

Jonathan Hyer, Haziq Raees Chowdhury, Richard Men Ho Lee, Syed Taseer Hasan

ABSTRACT

Introduction: Vertical gaze palsy is a highly relevant clinical sign in Parkinsonian syndromes. As the eponymous sign of progressive supranuclear palsy (PSP), it is a core feature of this disease. However, the differential diagnosis of vertical gaze palsy in patients with Parkinsonism is rarely considered.

Case Report: We present a case of a 65-year-old male with Parkinsonism who presented with diplopia secondary to oculomotor dysfunction and vertical gaze palsy. Computed tomography (CT) scan demonstrated a thalamic hemorrhage with intraventricular extension. Neurosurgical intervention was not required. The patient’s spectacles were fitted with a temporary Fresnel prism over the left lens to relieve diplopia. This progressively improved without the need for strabismus surgery.

Conclusion: Vertical gaze dysfunction occurs in patients with thalamic hemorrhage and is an important differential diagnosis of vertical gaze palsy in Parkinsonism.
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Keywords: Parkinson’s disease, Vertical gaze palsy, Progressive supranuclear palsy, Thalamic haemorrhage

INTRODUCTION

The differential diagnosis of Parkinsonism can be challenging. Despite consensus on diagnostic criteria for Parkinson’s disease and the various atypical Parkinsonian disorders including progressive supranuclear palsy (PSP), there is a high rate of mis-diagnosis particularly in the early stages [1]. Furthermore, there is no diagnostic laboratory or imaging test for these atypical disorders. Conventional magnetic resonance imaging (MRI) is insufficiently sensitive or specific for routine workup of PSP or other neurodegenerative Parkinsonian disorders and is compounded by high variability based on the experience of the neuroradiologist [2]. Considerable overlap may exist in the clinical presentation of atypical Parkinsonian disorders and other diseases and as such, presence of vertical gaze palsy is open to error as a predictor of pathology. The presence of vertical gaze palsy may lead to significant false positive diagnosis of PSP. Furthermore, a weakness in the vertical gaze is often observed in elderly patients. Vertical gaze palsy on a background of Parkinsonism strongly suggests PSP but also rarely occurs in other atypical Parkinsonian disorders such as diffuse Lewy body disease [3]. Clinically, vertical gaze palsy appears relatively selective and typically affects primary vertical saccades as well as vertical smooth eye movements to varying degrees [4].
Vertical ocular palsies are most commonly in up-gaze, less frequently are combined up- and down-gaze and rarely pure down-gaze. The nature of the palsy helps define both clinical and anatomical diagnosis [5]. It is important to consider and exclude vascular lesions, neoplasia, demyelination and infection for which brain imaging can be particularly useful. Accurate and timely diagnosis of these differentials is imperative both therapeutically and prognostically.

CASE REPORT

A 65-year-old male with Parkinsonism presented to ophthalmology casualty with a two-day history of binocular diplopia and unsteadiness following an episode of dizziness three days previously. He was otherwise well in himself with no other neurological symptoms and in particular, no headache. There was no history of trauma. There was no significant past medical history and his only medication was levodopa. There was no past notable ocular history. On examination, uncorrected visual acuity was 6/12 in both eyes. Bilateral ptosis was noted. Examination of eye movements identified a vertical gaze palsy affecting up-gaze saccades and pursuit. There was a slight left sided exo-tropia and hypotropia in the primary position with mild weakness of medial rectus. There was no nystagmus. Pupil responses were equal and reactive to light and there was no relative afferent pupillary defect. Color vision was full on Ishihara plate testing and exophthalmometry revealed no proptosis. Dilated fundal examination was normal with a healthy optic disc appearance. Intraocular pressures were also normal. Blood pressure was 123/84 mmHg and random blood glucose was 5.3 mmol/L. Cardiovascular examination was normal with no heart murmurs or carotid bruits on auscultation.

Differential diagnosis prior to neuroimaging was that of an atypical Parkinsonian disorder such as PSP, multisystem atrophy, corticobasal degeneration and diffuse Lewy body disease. Other common causes of vertical gaze palsies were considered such as thalamic or brainstem stroke, neoplasm or hydrocephalus. Lack of pupil involvement and convergence nystagmus was against a diagnosis of Parinaud syndrome. Medical causes of third nerve palsy were also considered.

Computed tomography (CT) scan identified a thalamic bleed with intraventricular extension (Figure 1). MRI scan confirmed a focal bleed in the thalamus posteriorly in close proximity to the pulvinar nucleus projecting into the retrothalamic cistern (Figure 2). The radiographic findings suggested a hypertensive or other vasculopathic focus rather than an aneurysm.

The patient was admitted and kept under neurological observation. No neurosurgical intervention was required. His spectacles were fitted with a temporary Fresnel prism over the left lens to relieve diplopia which gradually resolved over the following two weeks with improvement in vertical gaze palsy (Figure 3). No strabismus surgery was required.
DISCUSSION

Acute thalamic stroke is an important cause of vertical gaze palsy and is common after thalamic hemorrhage, especially associated with intraventricular extension [6]. While acute thalamic infarction appears to carry an overall good prognosis, thalamic hemorrhage is frequently associated with early neurological deterioration, severe deficits and high mortality and requires urgent neurosurgical referral [6, 7]. Computed tomography (CT) scan of head is the imaging procedure of choice and once the diagnosis is confirmed blood pressure should be maintained below a mean arterial pressure of 130 mmHg in a person with no history of hypertension [8]. Standard craniotomy for surgical removal of thalamic hemorrhages has been all but abandoned because of extremely poor outcomes in most patients. Angiography is not required for older hypertensive patients with a thalamic haemorrhage and in whom CT scan findings do not suggest a structural lesion.

There are no reports of vertical gaze palsy having occurred co-incidentally in a patient with Parkinsonism. Kumral et al. [6] evaluated a cohort of patients with pure thalamic ischaemic stroke and identified up-gaze palsy in 50% patients with bilateral paramedian infarction and 25% with unilateral paramedian infarction [6]. A case of unilateral thalamic infarction presenting with vertical gaze palsy and skew deviation was reported [9]. Reversibility of ocular manifestations of thalamic hemorrhage following ventricular drainage has also been described [10, 11].

The excellent outcome in our case is unusual. It has been suggested that the up-gaze palsy in a case of Papinaud Syndrome associated with thalamic hemorrhage may be attributed to increased intracranial pressure resulting from a mass effect on the pre-tectal region and tectum, or to tightness in the incisura causing hydrocephalus secondary to aqueduct compression [11]. This may be relevant to our patient and help explain the improvement in eye movements as the hemorrhage was reabsorbed. Our patient benefited from the use of a prismatic spectacle lens. A study investigating prismatic spectacle lenses on symptoms of dizziness, headache and anxiety caused by vertical heterophoria is currently ongoing [12].

CONCLUSION

Alternative diagnoses other than progressive supranuclear palsy in patients with Parkinsonism and vertical gaze palsy should be considered as it is also featured in patients with thalamic hemorrhages, which can be easily identified on brain imaging. All patients presenting with an intraventricular hemorrhage need careful blood pressure monitoring and urgent neurosurgical review.

Author Contributions
Jonathan Hyer – Conception and design, Drafting the article, Final approval of the version to be published
Haziq Raees Chowdhury – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Richard Men Ho Lee – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Syed Taseer Hasan – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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