



Parinaud Syndrome in Association with Thalamic Infarct in a Young Person

Priscilla Xinhui Wang^{1*} and Srinivasan Sanjay²

¹Department of Ophthalmology, National University Health System, Singapore.

²Department of Ophthalmology and Visual Sciences, Khoo Teck Puat Hospital, Yong Loo Lin School of Medicine, National University of Singapore, Singapore.

Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

Case Study

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ABSTRACT

Aim: We report a case of Parinaud syndrome secondary to an acute left posteromedial thalamic infarct in a young person.

Presentation of Case: Patient's main presenting complaints were blurring of vision and diplopia. The main clinical manifestations were that of a severe restriction in upward gaze, and convergence-retraction nystagmus.

Discussion: Vertical gaze palsies have been well associated with midbrain lesions but more rarely associated with seemingly isolated thalamic lesions. This case report further supports current limited literature that suggests that thalamic lesions may also manifest as vertical gaze palsies. In addition, cases of cerebrovascular events are usually first seen in the Emergency Department where diagnoses are made and acute management is given. In this case, the patient was referred to the Eye clinic from the Emergency Department for visual symptoms. Parinaud syndrome was promptly recognised by the ophthalmologist upon examination and the decision for urgent brain imaging with referral to the neurologist was quickly made. Timely management of patient likely contributed to his favourable outcome.

Conclusion: This is a case of Parinaud syndrome secondary to an acute left posteromedial thalamic infarct in a young person without obvious midbrain involvement. This case highlights the important role of ophthalmologists in the early recognition of Parinaud syndrome, which can allow for prompt diagnosis and management. Timely diagnosis of Parinaud syndrome can be crucial to patient's outcome.

*Corresponding author: E-mail: priscilla.wang@mohh.com.sg;

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1. INTRODUCTION

Parinaud syndrome, now better known as dorsal midbrain syndrome, is a supranuclear paralysis of the vertical gaze resulting from damage to the mesencephalic tectum. It consists of abnormalities of eye movement and pupil dysfunction, namely vertical gaze palsies, convergence-retraction nystagmus, convergence bias with horizontal saccades, light-near dissociation, corectopia, spastic-paretic accommodation and pathological lid retraction. It results from direct or compressive injury to the dorsal midbrain, and can arise from a variety of causes, such as pineal, midbrain or 3rd ventricle tumors, aqueductal stenosis, multiple sclerosis, arteriovenous malformations, encephalitis, obstructive hydrocephalus, uncus herniation, and cerebrovascular disorders [1-11]. While lesions of the mesencephalic reticular formation are known to produce vertical gaze palsies, herein, we report an interesting case of Parinaud syndrome which manifested in a young patient with thalamic infarction.

2. CASE REPORT

We report an interesting case of a 35-year-old Indian male who was referred by the Emergency Department to the Department Of Ophthalmology and Visual Sciences, Khoo Teck Puat Hospital, Singapore, for an acute onset of bilateral painless blurring of vision associated with diplopia of 3-day duration. His symptoms first started when he was seated down and having his breakfast. They were persistent and did not improve or worsen with time. He had no history of trauma or fall. There was no associated focal numbness or weakness, headache, nausea or vomiting. The patient was a smoker of 8.5 pack years, and was diagnosed with diabetes mellitus type 2 three years ago. He was followed up by a private general practitioner and was taking oral hypoglycemic agents. However, there was no record of his glycaemic control available and patient was also not sure of his diabetic control. He denied previous hypoglycemic episodes. He had no other known ischemic risk factors such as hypertension, ischemic heart disease or hyperlipidemia, and no prior ocular history.

On examination, distant Snellen visual acuity was 6/15 for both eyes, best corrected with pinhole to 6/12 in the right eye and 6/9 in the left eye. Refraction was not performed. Patient did not have a history of wearing glasses or contact lens. Direct and consensual pupillary light reflexes were normal. There was no relative afferent pupillary defect. His lenses were clear. Intraocular pressure was normal. Anterior and posterior segments were unremarkable. However, patient had a severe restriction in upward gaze and a mild restriction in downward gaze. There was also convergence-retraction nystagmus on attempted upward gaze. Doll's head maneuver elevated the eyes. Eye movements were otherwise full in lateral gazes. Binocular vertical diplopia was present for both far and near vision. There was convergence bias with horizontal saccades. He did not have light-near dissociation, corectopia or Collier's sign. Bell's phenomenon and skew deviation were not investigated. Fig. 1. shows the clinical pictures of patient's eyes in different directions of gazes. However, we recognize that the data shown in pictures do not always correlate with the clinical examination.

On further examination, patient had normal cranial nerve function, full power in all limbs, and intact sensation. However, he had unsteady gait and was unable to perform tandem gait. Patient had complained that he was feeling too giddy to do so. Glasgow Coma Scale was 15.

Based on these findings, a provisional diagnosis of Parinaud syndrome was made. Possible causes included brain tumors in the pineal gland, such as intracranial germinomas, as well as multiple sclerosis, arteriovenous malformation and stroke. Patient was admitted on the same day and magnetic resonance imaging (MRI) and angiography of the brain confirmed an acute infarct in the left posteromedial thalamus, extending slightly into the left cerebral peduncle. Figs. 2, 3 and 4 show various cuts of the MRI performed. A neurologist further confirmed the diagnosis. The patient's systolic blood pressure was noted to range from 100 to 120mmHg while his diastolic blood pressure had ranged from 70 to 80mmHg. Lipid panel revealed total cholesterol of 5.13mmol/L, triglycerides of 1.92mmol/L, high-density lipoprotein (HDL) of 0.81mmol/L, low-density lipoprotein (LDL) of 3.88mmol/L, and a cholesterol to HDL ratio of 6.33, which essentially put the patient at a high risk for cardiovascular diseases. His diabetic control was unacceptable with a glycated HB of 9.2% (77mmol/mol). Thyroid, liver and renal function tests were unremarkable. Calcium panel and magnesium level were within normal range. Full blood counts, peripheral blood film, prothrombin time, international normalized ratio, and activated partial thromboplastin time were normal. A young stroke work-up comprising of anti-nuclear antibody, anti-double-stranded-DNA antibody, lupus anticoagulant and anti-cardiolipin antibodies were negative. Homocysteine, anti-thrombin III, activity profiles of protein C and S were normal too. Ultrasound Doppler of carotid arteries was unremarkable.

Patient was started on aspirin 100mg once every morning, and simvastatin 20mg once every night. To improve control of his diabetes, physicians made adjustments to his oral hypoglycemic agents, and provided dietary and lifestyle advice. Patient was also given counselling on smoking cessation.

Over the course of his 4-day inpatient stay, patient experienced symptomatic and clinical improvement, with resolving giddiness and diplopia as his restriction in upward gaze improved. He was subsequently able to ambulate well prior to discharge.



Fig. 1. Shows the clinical pictures of patient's eyes in different directions of gazes. Figure 1b shows a severe restriction in upward gaze while Figure 1h shows a mild restriction in downward gaze

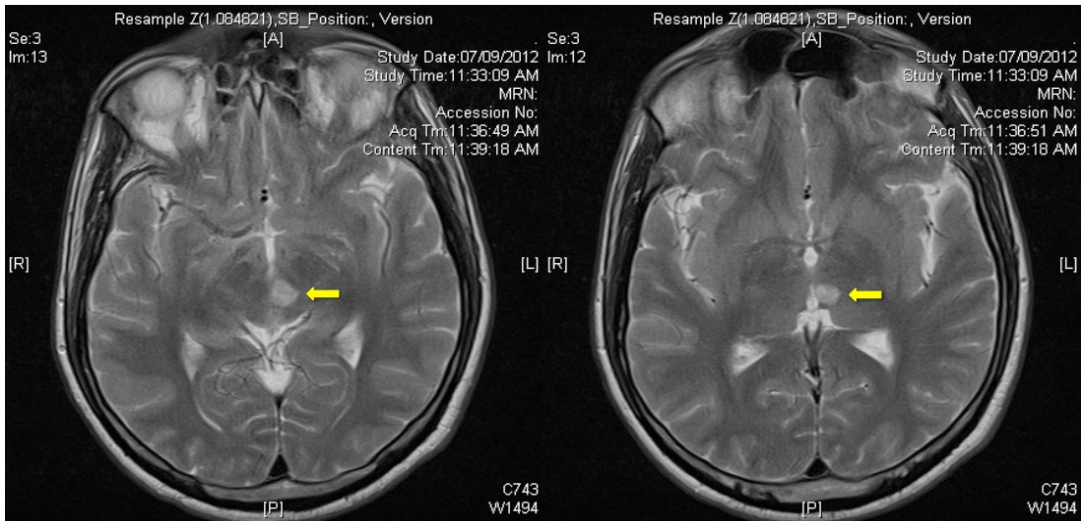


Fig. 2. Shows T2-weighted MRI of the patient's brain in axial cuts. The yellow arrows point to the acute left posteromedial thalamic infarct

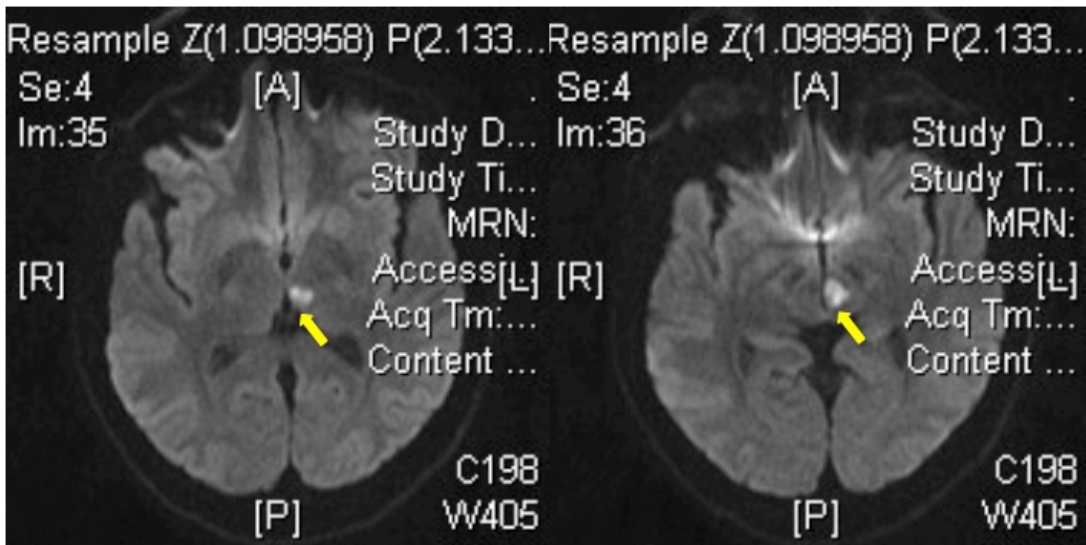


Fig. 3. Shows diffusion MRI of the patient's brain in axial cuts. The yellow arrows point to the acute left posteromedial thalamic infarct

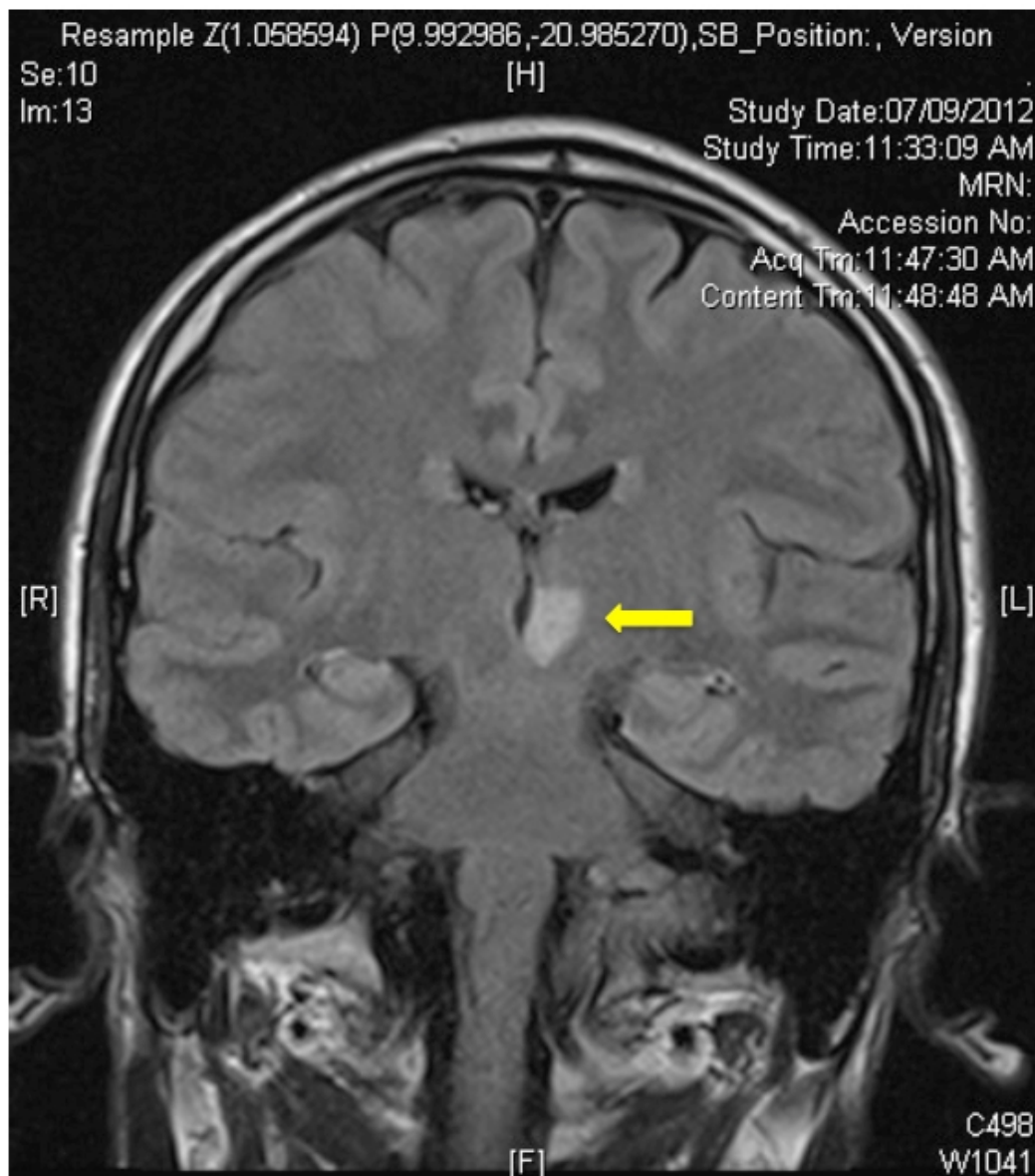


Fig. 4. Shows T2 FLAIR MRI of the patient's brain in a coronal cut. The yellow arrow points to the acute left medial thalamic infarct

3. DISCUSSION

Vertical gaze palsy has been reported to be caused by lesions involving bilateral rostral interstitial nucleus of medial longitudinal fasciculus (riMLF) or unilateral posterior commissure, while convergence nystagmus is usually caused by a lesion near or within the dorsal mesencephalon [12,13]. Later reports demonstrated that vertical gaze palsy and convergence nystagmus can result from unilateral riMLF lesion [14-17]. Each riMLF contain burst neurons for both upward and downward movements [18]. A unilateral lesion of the

riMLF may occasionally cause combined upgaze and downgaze palsies, perhaps by disrupting bilateral upward gaze excitatory and inhibitory inputs and unilateral downgaze excitatory inputs [19]. Fibres responsible for upward gaze cross rostrally and posteriorly between the riMLF nuclei (near the pineal body) through the posterior commissure and are subject to interruption before descending and terminating in the contralateral oculomotor nuclei. On the other hand, pathways for downward gaze project directly downward from the riMLF to the oculomotor complex. This may be a reason for the dissociation of the upward and downward gaze palsies, and the reason why upward gaze appears to be affected more than downward gaze [20]. There have been a few anatomic-clinical cases which demonstrated that supranuclear palsies of the downward gaze were related to paramedian lesions of the rostral mesencephalon involving the riMLF, the nucleus of Cajal, and/or their afferent and/or efferent pathways, while supranuclear palsy of the upward gaze was related to lesions of the posterior commissure [12].

Lesions of the mesencephalic reticular formation including the nucleus of Darkschewitsch, the rostral interstitial medial longitudinal fasciculus, the interstitial nucleus of Cajal, and the posterior commissure, are known to produce vertical gaze palsies [21]. In general, vertical gaze palsies have been attributed to associated lesions of vertical eye movement control centres in the rostral midbrain rather than the thalamic injury. The frequent coexistence of both midbrain and paramedian thalamic infarct is related to their vascular supply; a single vessel arising near the top of the basilar may branch to supply both the paramedian region of the thalamus and the rostral medial mesencephalon [22]. There have been only a few cases with MRI-documented medial thalamic infarctions without midbrain involvement who presented with vertical gaze palsies [21,23]. In our case, patient had vertical gaze palsy and was found to have an acute infarct in the left medial thalamus. There was no obvious midbrain involvement on the imaging studies. It could be that microinfarcts in the dorsal mesencephalic structures sharing a common vascular supply were too small to be detected [21,24], or that the supranuclear inputs were interrupted [21], possibly by the surrounding edema resulting from the infarct. Perhaps serial brain imaging done over a space of time could have provided more information.

Convergence-retraction nystagmus is believed to be a saccadic disorder rather a nystagmus because the primary adductive movements are asynchronous adducting saccades [25]. It is not a true nystagmus because there is no slow phase. Convergence and globe retraction occurs when attempting upward saccades. Limitation of upward gaze as well as defective upward saccades and pursuit are always present. Globe retraction occurs as a result of co-contraction of the medial rectus muscles. It is often best elicited by down-moving optokinetic nystagmus stimulus since upward movement of the eyes is replaced by convergence retraction movement. Convergence-retraction nystagmus has been reported as a classic finding in extensive and bilateral lesions of the dorsal midbrain, which are usually neoplastic and signal poor prognosis [26]. However, in our case, while the patient did have convergence-retraction nystagmus, he did not have extensive or bilateral lesions of the dorsal midbrain and had experienced instead, a good functional recovery. Doll's head maneuver could elevate the patient's eyes as it was a supranuclear vertical gaze palsy. In this case, patient's Bell's phenomenon would have been preserved since there was no lower motor neuron palsy. The patient was having diplopia possibly from a skew deviation, or a subtle hypertropia or hyperphoria. Unfortunately, these signs were not investigated and picked up clinically.

Benign course with favourable outcome has been reported for cerebrovascular events with conservative management [26,27]. On the other hand, surgical treatment such as

transpositions and inferior rectus recessions have also demonstrated improvement in upgaze limitation and convergence-retraction nystagmus [28]. Parinaud syndrome associated with brain tumors in pineal gland or midbrain is seen more frequently in younger patients, while in older patients, it is more commonly associated with cerebrovascular disorders such as hemorrhage, ischemia and infarct. In our case study, although the patient was young, he had a few ischemic risk factors such as diabetes and a smoking history. Hyperlipidemia was only detected after investigations. He had no autoimmune or thrombophilic disorders. He experienced symptomatic and clinical improvement with non-invasive and conservative management.

Prognosis for Parinaud syndrome depends on the causative factor. Parinaud syndrome has been classically associated with young patients with brain tumors in the pineal gland or midbrain, with intracranial germinomas being the most common lesion. Generally, intracranial germinomas have excellent prognosis due to their sensitivity to radiotherapy [29]. Parinaud syndrome can also manifest in patients with multiple sclerosis, or following stroke of the upper brainstem, like the patient in this case. Factors that led to his favourable outcome likely included his young age, lack of existing co-morbidities and the fact that the infarct was not extensive. Patient's long-term outcome would likely be dependent on his control of his cardiovascular risk factors.

Cases of cerebrovascular events are usually first seen by the Emergency Department where diagnoses are made and acute management is given. In this case, the patient was referred to the Eye clinic from the Emergency Department for visual symptoms. Parinaud syndrome was promptly recognised by the ophthalmologist upon examination and the decision for urgent brain imaging with referral to the neurologist was quickly made. Timely management of patient likely contributed to his favourable outcome. Patients presenting with symptoms such as vertical gaze palsies and convergence-retraction nystagmus should prompt physicians to perform neuroimaging studies quickly, especially if risk factors for cerebrovascular events are present.

4. CONCLUSION

This is a case of Parinaud syndrome secondary to an acute left posteromedial thalamic infarct which extended slightly into the left cerebral peduncle. This further supports current literature which suggests that Parinaud syndrome can manifest in thalamic infarctions even without obvious midbrain involvement. Patient's main presenting complaint was diplopia with blurring of vision, and the main clinical manifestations were that of a severe restriction in upward gaze, and convergence-retraction nystagmus. This case highlights the important role of ophthalmologists in the early recognition of Parinaud syndrome, as it may be the sole manifestation of cerebrovascular events. Early recognition can allow for timely referral and prompt imaging for diagnosis and subsequent management. This can be crucial to patients' outcome.

CONSENT

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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