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Is there variable neuronal susceptibility to compression? A case report of differential recovery from dorsal midbrain syndrome and compressive optic neuropathy following treatment for germinoma

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Abstract

A 12-year-old boy with suprasellar germinoma complicated by hydrocephalus was found to have dorsal midbrain syndrome and bilateral compressive optic neuropathy. Following a third ventricular neuroendoscopic biopsy and the initiation of chemotherapy, a follow-up magnetic resonance imaging scan of the brain showed significant regression of the tumor with resolution of hydrocephalus. On smooth pursuit, there was significant improvement in upgaze. However, light-near dissociation of his pupillary defect was persistent. Fundal examination showed persistent temporal pallor of the optic discs bilaterally, associated with a suboptimal best-corrected visual acuity of 20/40 and bilateral centrocecal scotoma.

The differential recovery of neuronal function following relief of compression is poorly understood. In this case, both the optic nerve and pretectal axonal fibers responsible for pupillary reflex, with their neuronal cell bodies residing outside the direct compressive site at the midbrain, showed poor functional recovery. However, relief of pressure at the rostral interstitial medial longitudinal fasciculus and posterior commissure nuclear complexes resulted in significant clinical improvement in upgaze.

This case suggests that, for neurons, axons are apparently more susceptible to pressure than cell bodies. The predominant transmission of nerve impulses takes place through the length of axons, and thus is more sensitive to pressure changes, with the associated microvascular compromise and demyelination.

Keywords: *hydrocephalus, germinoma, optic nerve diseases, Parinaud syndrome*

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Introduction

Of all germinomas which occur in the central nervous system, 25%-35% are found in the suprasellar region.[1] Obstruction of the flow of cerebrospinal fluid, with subsequent hydrocephalus and dilatation of the aqueduct of Sylvia by the tumour is a known cause of dorsal midbrain syndrome.[2]

Although it is generally believed that neurons of the central nervous system have limited regenerative capacity, patients with this condition may improve following the treatment for hydrocephalus.[3] However, we observed that not all neurological deficits resulting from hydrocephalus recover to the same extent following treatment. Even within the constellation of clinical signs comprising the dorsal midbrain syndrome, some features may resolve better than the others. In this case report, we described the evolution of clinical signs following the treatment of a suprasellar germinoma.

Case presentation

A 12-year-old boy with good past health complained of polyuria and polydipsia for one month. On blood tests he was noted to have normal blood glucose but hypernatremia suggestive of diabetes insipidus.

Magnetic resonance imaging (MRI) of the brain showed a 2.2 x 1.9 x 1.5 cm T1W isointense, T2W hyperintense, echancing mass at the suprasellar cistern and hypothalamus. Contrast-enhancing lesions were also detected at the pineal region and walls of the third ventricle, leading to hydrocephalus (Fig. 1).

Ophthalmic examination showed features of dorsal midbrain syndrome with failure of upgaze (Fig. 2), convergence-retraction nystagmus, and light-near dissociation of pupillary reaction (Fig. 3). Convergence was full. He also had mild temporal pallor of the optic discs associated with centrocecal scotoma bilaterally. His best-corrected visual acuity was 20/40 in both eyes. The function of other cranial nerves was intact. Limited by the schedule of his systemic treatment, optical coherence tomography scan of the retinal nerve fibre layer could not be arranged.

The pathology of the mass was confirmed to be germinoma during a third ventricular neuroendoscopic biopsy. Following hormonal replacement for panhypopituitarism and cranial diabetes insipidus, the patient underwent chemotherapy with etoposide and cisplatin.

Four weeks later, a follow-up MRI scan of the brain showed significant regression of the multifocal tumours with resolution of hydrocephalus (Fig. 4). His ocular motility showed significant improvement. On smooth pursuit, there was no obvious limitation in upgaze (Fig. 5). Only minimal limitation was noticed on upward saccade. However, his pupillary light-near dissociation was persistent (Fig. 6). The patient's best-corrected visual acuity remained suboptimal at 20/40

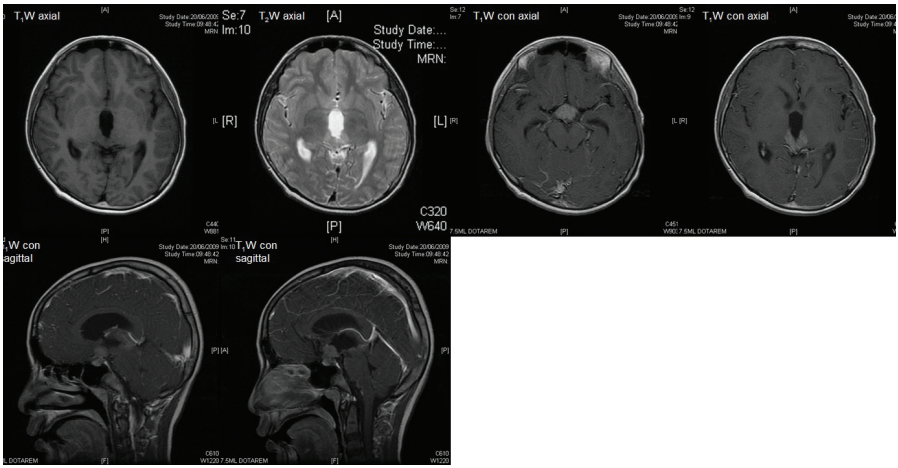

Fig. 1. Magnetic resonance imaging of the brain showed a 2.2 x 1.9 x 1.5 cm mass at the suprasellar cistern and hypothalamus which is T1W isointense (upper panel, 1st picture from the left), T2W hyperintense (upper panel, 2nd picture from the left), and contrast-enhancing (upper panel, 3rd picture from the left). Contrast-enhancing lesions were also detected at the pineal region and walls of the third ventricle, leading to hydrocephalus (upper panel, 4th picture from the left). T2W contrast sagittal cuts (lower panel) revealed the enhancing suprasellar mass and multiple enhancing lesions along the cervical spinal cord, suggestive of metastasis. The aqueduct of Sylvia was grossly dilated, causing dorsal midbrain syndrome.

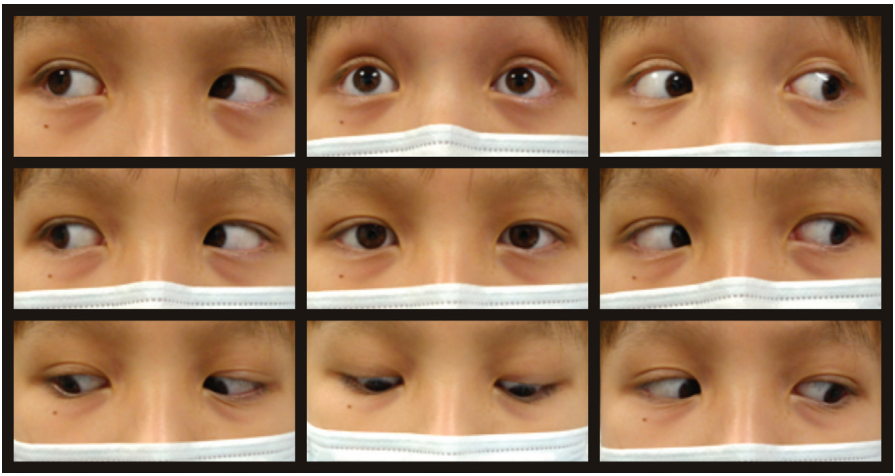

Fig. 2. Nine gaze photos of the patient before treatment. He had profound limitation in upgaze.

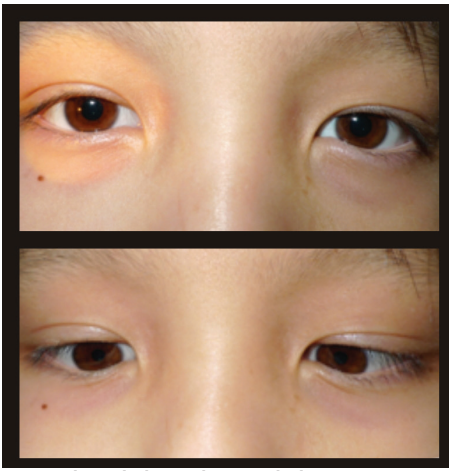

Fig. 3. Clinical photo showing light-near dissociation of the patient's pupillary reflex before treatment. Note the dilated pupil as the torch light was shone onto the patient's right eye (upper picture). The pupils constricted as he accommodated for near objects (lower picture).

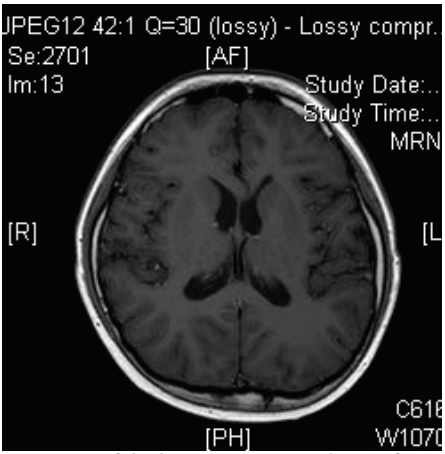

Fig. 4. MRI of the brain (T1W coronal view) four weeks after third ventricular neuroendoscopic biopsy and drainage, and the initiation of chemotherapy. Both the suprasellar tumor and hydrocephalus had resolved.

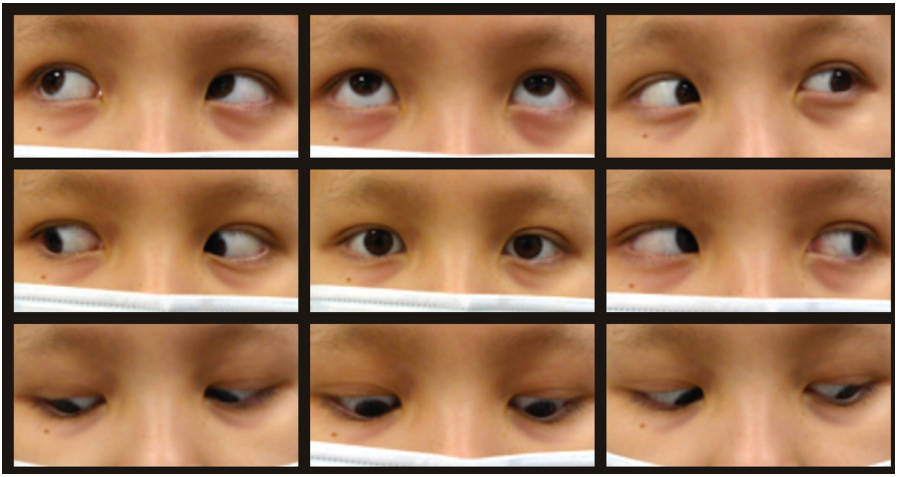

Fig. 5. Nine gaze photos of the patient four weeks after third ventricular neuroendoscopic biopsy and drainage, and the initiation of chemotherapy. There was significant improvement in upgaze. The patient lost his hair as a side effect of the chemotherapy.

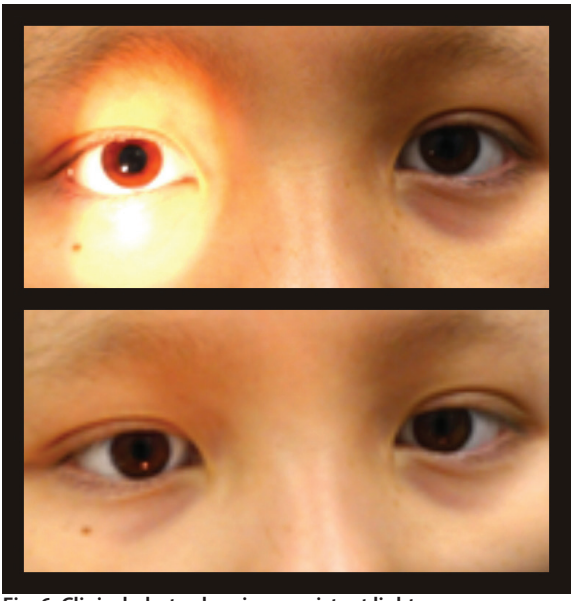

Fig. 6. Clinical photo showing persistent light-near dissociation of the patient's pupillary reflex after treatment. Note the dilated pupil as the torch light was shone onto the patient's right eye (upper picture). The pupils constricted as he accommodated for near objects (lower picture).

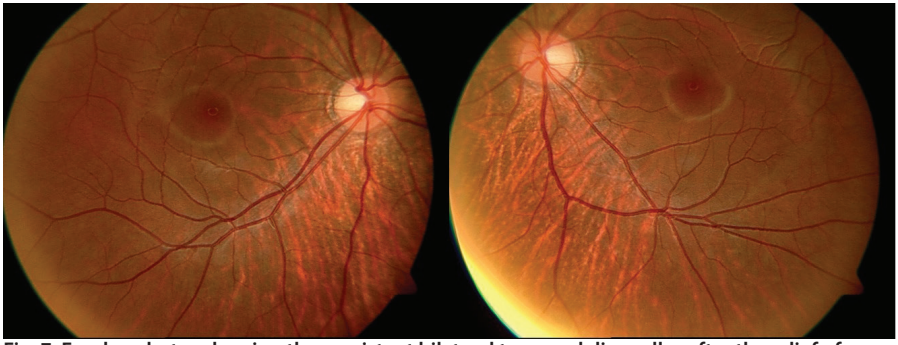

Fig. 7. Fundus photos showing the persistent bilateral temporal disc pallor after the relief of hydrocephalus with treatment.

in both eyes, and the associated centrocecal scotoma was persistent. Fundal examination showed persistent temporal pallor of the optic discs bilaterally (Fig. 7).

Discussion

In 1883, Henri Parinaud described a clinical syndrome consisting of vertical gaze paralysis, convergence-retraction nystagmus on attempted upgaze, light-near dissociation of pupillary reflex, lid retraction (Collier's sign), and skew deviation.[4] Known as the dorsal midbrain syndrome, clinical-anatomical correlation has been made in its constituent features. The upgaze palsy has been attributed to the increased size of the aqueduct producing both mechanical stretch and compromise to the blood supply to the rostral interstitial nucleus of medial longitudinal fasciculus (riMLF) and its connection to the ventral posterior commissure. On the other hand, dysfunction of the brachium of the superior colliculus and pre-tecto-oculomotor fibres has been speculated as the cause of light-near dissociation of pupillary reflex.[5]

The association between hydrocephalus and dorsal midbrain syndrome has been well established.[6] In the interventional case series by Bergsneider *et al.*, it was found that clinical manifestations of dorsal midbrain syndrome could be reversed once the intracranial pressure and ventricular size were normalized by siphoning.[7]

This case report is the first to describe that the clinical features of dorsal midbrain syndrome can resolve to different extents following treatment of the compressive lesion. In this case, significant recovery was observed in upgaze but not light-near dissociation of pupillary reflex. In the presence of concurrent compressive optic neuropathy, no significant recovery in optic nerve function was demonstrated following the relief of hydrocephalus.

The differential recovery of neuronal function following relief of compression is poorly understood. In this case, both the optic nerve and pretectal axonal fibres responsible for pupillary reflex, with their neuronal cell bodies residing outside the direct compressive site at the midbrain, showed poor functional recovery. On the contrary, relief of pressure at the riMLF and posterior commissure nuclear complexes resulted in significant clinical improvement in upgaze.

This case suggests that for neurons, axons are apparently more susceptible to pressure than cell bodies. The predominant transmission of nerve impulses takes place through the length of axons, and thus is more sensitive to pressure changes, with the associated microvascular compromise and demyelination.

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