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LETTER TO THE EDITOR

Treatment of normal pressure hydrocephalus resolves dorsal midbrain syndrome

Joshua A. Smith[1] · Fahad J. Laghari[1] · Sandra M. Pinzon[1]

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Dear Editors,

We present a unique case of a patient presenting with the signs and symptoms of normal pressure hydrocephalus (NPH) and dorsal midbrain syndrome (DMS), and in whom treatment of former led to the improvement of both. DMS has often been described in the presence of increased intracranial pressure and hydrocephalus; however, according to our knowledge, this is the first report that describes a relationship between NPH and DMS.

A patient in his seventies was admitted to the hospital under the medicine service for pneumonia, sepsis, and falls. The CT scan of the head showed dilated ventricles out of proportion to sulcal size, concerning for NPH (Fig. 1). Neurology was consulted for confusion, falls, and abnormal CT scan of the head. The patient had been started on memantine by a primary care physician for almost a year. A formal inpatient mental status test was not performed as the patient's confusion may have been exaggerated by sepsis, and the records from PCP were unavailable. He had slowly developed gait dysfunction over the past months and felt as if his feet were stuck to the floor. The patient had also developed incontinence recently.

On examination, the patient had cognitive slowing and difficulty with short-term recall, although recent and remote memory appeared to be intact. During physical examination, the patient also had upgaze paresis and was unable to perform accommodation. He walked stooped forward, legs shoulder width apart, took small steps with occasional festination, and at times, initiation of movement was slowed. MRI of the head was completed to rule out other compressive lesions which could lead to both obstructive hydrocephalus and DMS. MRI did not reveal obstructive hydrocephalus. Interestingly, there was compression of the midbrain by the dilated third ventricle (Fig. 2). The diagnosis of NPH and DMS was deduced based on history, physical examination, and the imaging.

The patient had a lumbar puncture (LP) performed revealing only a mild increase in opening pressure, 23 mmHg. Patient showed improvement in gait speed following 40 mL CSF drain. Later, patient received a trial of lumbar drain which showed improvement in dynamic gait index from 2 to 9 in a single day, and the decision for ventriculoperitoneal shunt (VPS) placement was made.

Subsequent evaluations following the VPS placement showed patient did not have vertical gaze palsy. It is reasonable to presume that the patient's vertical gaze palsy and thus his DMS improved, if not resolved, with shunting.

DMS is also known as Parinaud syndrome, Sylvian aqueduct syndrome, pretectal syndrome, and Koerber-Salus-

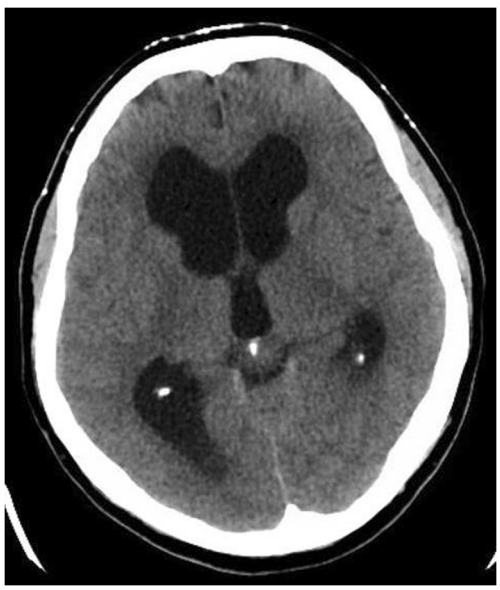

Fig. 1 CT head axial cut revealing ventriculomegaly out of proportion to sulcal size. Lateral ventricle as well as third ventricle enlargement can be seen on this image

✉ Fahad J. Laghari
laghariffj2@upmc.edu

[1] Department of Neurology, University of Pittsburgh Medical Center Hamot, 201 State Street, Erie, PA 16550, USA

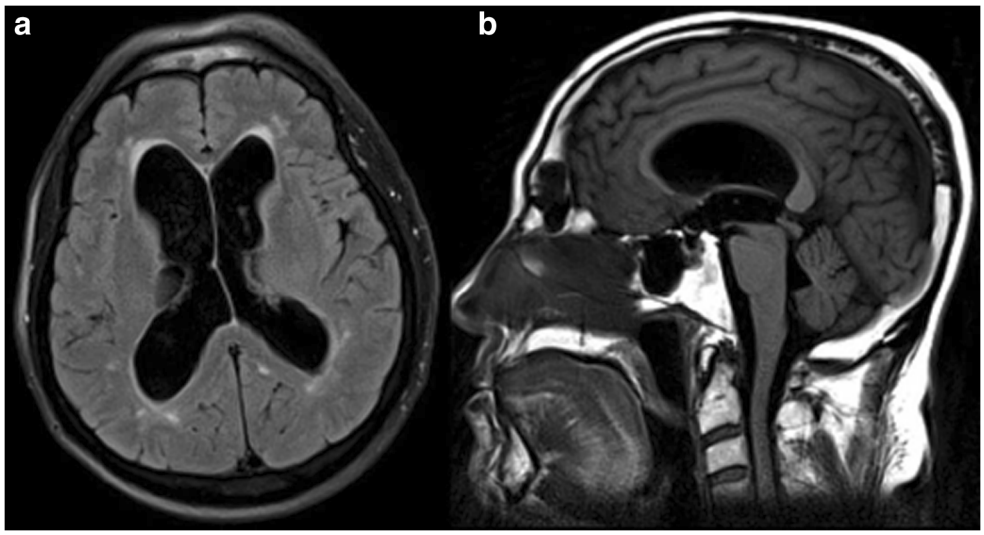

Fig. 2 **a** MRI FLAIR axial cut revealing ventriculomegaly out of proportion to sulcal size as well as T2 hyperintensities in the white matter. **b** MRI T1 sagittal cut revealing midbrain compression by a dilated third ventricle

Elsching syndrome. It consists of a group of signs found in those patients who have conditions affecting the dorsal part of the midbrain or the pretectal region especially the rostral interstitial nuclei of the medial longitudinal fasciculus (riMLF) [1]. The core signs include paralysis of the upward gaze and convergence. Patients may also have mild mydriasis, convergence retraction nystagmus, lid retraction, and ptosis. Often, the patients also have the light-near dissociation and mid-sized pupils (4–6 mm) [2].

A literature search reveals that hydrocephalus with increased intracranial pressure, ischemic strokes, multiple sclerosis, and tumors are the common causes of the DMS [3]. It has been postulated that the symptoms of DMS in patients with hydrocephalus occur as a result of increased intracranial pressure, as relieving the pressure relieves the ocular symptoms [4]. Furthermore, in patients with shunted hydrocephalus, signs of DMS may be an indicator of increased intracranial pressure and shunt failure [5].

The sagittal MRI image (Fig. 2b) may appear to show midbrain atrophy and "colibri sign" which, along with patient's gait, could represent progressive supranuclear palsy (PSP). PSP is a neurodegenerative disease that presents in elderly individuals, usually in their sixties and seventies, with parkinsonian symptoms and extraocular movement palsies. There have been reports of patients with neurodegenerative diseases that have been misdiagnosed as NPH [6]. It is unlikely that a patient suffering from PSP presents with symptoms of gait dysfunction, cognitive decline, urinary incontinence, and upward gaze paresis that significantly improve with a VP shunt. The patient also did not have rigidity, lacked bradykinesia apart from lower extremities, was not falling backward, and their pons on MRI did not appear spared. The latter finding goes on to show that the midbrain and pons findings are probably due to compression by an enlarged third ventricle instead of the neuronal loss in the midbrain as observed in PSP.

DMS has been previously described in patients with hydrocephalus with increased intracranial pressure but not with NPH. Our case report describes a patient with NPH with normal-high intracranial pressure and signs of DMS. The MRI of the patient showed ventriculomegaly and conformational distortion of the dorsal midbrain. Patients with NPH often present with a triad of slowly progressive gait disturbance, impairment of cognitive function, and urinary incontinence. The patient described in this case report underwent treatment for NPH: initially with lumbar punctures and ultimately with VPS leading to resolution of signs and symptoms of both the DMS and NPH. This case report adds evidence to the hypothesis that DMS, in hydrocephalus, may result from distortion of the pretectal area from enlargement of the third ventricle.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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