CASE REPORT

Dorsal Midbrain Syndrome from Thalamocapsuloganglionic Hemorrhage: A Case Report

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ABSTRACT

Dorsal midbrain syndrome (DMS) is a supranuclear palsy of vertical gaze characterized by conjugate upgaze palsy, light-near dissociation, convergence-retraction nystagmus, lid retraction, and skew deviation. Majority of cases are due to primary midbrain lesions such as strokes or neoplasms, or due to pineal gland tumors compressing the said area. Presented here is the case of a 57-year-old male who came in with a chief complaint of diplopia and the typical signs of DMS. Cranial tomography scan revealed a parenchymal hemorrhage at the left thalamocapsuloganglionic region, a rarely reported site of a primary lesion causing DMS. In this case, the syndrome may have been a consequence of the mass effects and perilesional edema associated with the thalamocapsuloganglionic hemorrhage, or may have been due to disruption of supranuclear inputs to the dorsal mibdrain. This case provides further evidence that DMS may arise from lesions without obvious involvement of the said region. This case also highlights the importance of a thorough physical examination to elicit the findings associated with DMS, and the need to correlate these with a keen analysis of diagnostic test results.

Keywords: dorsal midbrain syndrome, Parinaud syndrome, conjugate gaze spasm, convergence insufficiency, skew deviation, thalamic hemorrhage

INTRODUCTION

Dorsal midbrain syndrome (DMS) pertains to a paralysis of vertical gaze secondary to damage to the pretectum, an isthmus between the superior colliculi and the thalamus.[1] Also known as pretectal syndrome, Sylvian aqueduct syndrome, or Parinaud syndrome, the condition was first described in the late 1800s by French ophthalmologist Henri Parinaud. Parinaud hypothesized it to be caused by a lesion of the quadrigeminal area, which then caused upgaze palsy and convergence paralysis.[2] The current understanding of DMS today, however, has been widened to include vertical gaze palsy, convergence-retraction nystagmus, light-near dissociation, convergence palsy, skew deviation, pathologic lid retraction, and increased square wave jerks.[1] The triad of upgaze palsy, convergence retraction nystagmus, and light-near dissociation is found in up to 65% of patients.[2]

Direct or compressive injury to the dorsal midbrain may be secondary to a variety of causes, including pineal, midbrain, or third ventricle tumors, multiple sclerosis, aqueductal stenosis, encephalitis, arteriovenous malformations, obstructive hydrocephalus, and cerebrovascular disorders.[3] This report focuses on an unusual case of DMS resulting from a thalamocapsuloganglionic hemorrhage. Located above the midbrain, this implicated region includes the thalamus,

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and the paired white matter tracts (internal capsules) and subcortical nuclei (basal ganglia) lateral to it.

CASE PRESENTATION

This is the case of a 57-year-old Filipino male who came in with a chief complaint of binocular, vertical diplopia at the outpatient clinic of the Department of Ophthalmology and Visual Sciences, Philippine General Hospital (PGH), Manila. His symptoms started four months prior when he experienced sudden-onset headache and subsequent loss of consciousness while at a meeting, prompting immediate consult at a provincial hospital. There was no history of fall or head trauma. The patient was a known hypertensive for 15 years, and allegedly had an episode of cerebral infarction 10 years prior. He was maintained on telmisartan, amlodipine, and carvedilol with good compliance to medications. The patient had no other known comorbidities and denied having any vices.

Upon admission at the local hospital, a plain cranial computed tomography (CT) scan was done which allegedly revealed an intraparenchymal hemorrhage at the left thalamocapsular region. The patient was monitored for 7 days and then discharged. In the interim, the patient experienced occasional episodes of diplopia and blurring of vision, but he did not return to the local hospital. The patient procured spectacles from a local optical shop but symptoms did not improve.

At the time of examination at the PGH, visual acuity at distance was 20/20 (logMAR 0.0) for both eyes, with a near visual acuity of Jaeger 5. Both eyes presented with mild lid retraction, but were otherwise grossly normal. Pupillary examination revealed both pupils at 3 mm and sluggishly reactive to light. On primary gaze, there was a slight left hypertropia. The patient had severe restriction on upgaze of both eyes, with noted binocular vertical diplopia on near and far vision (Figure 1). Extraocular movements were normal in all other cardinal gazes, but the patient presented with skew deviation on further examination (Figure 2). On testing the near response, the patient had convergence-retraction nystagmus and light-near dissociation. Slit lamp examination was essentially unremarkable, save for immature cataracts in both eyes. Posterior pole exam revealed pink-orange discs with a cup-disc ratio of 0.6 in both eyes, with no apparent signs of hypertensive retinopathy.

Cranial nerve examination revealed a shallow nasolabial fold and subjectively decreased gross hearing on the right side. The patient had full strength in all limbs with intact sensation, but was slightly slow to walk. Glasgow Coma Scale score was at 15.

Based on the history and physical examination, a diagnosis of DMS was made. The patient was advised to retrieve his CT scan results and to follow up them. Review of the CT scan revealed a 5.4 cc parenchymal hemorrhage in the left thalamocapsuloganglionic region, with associated compression of the body and posterior horn of the left lateral

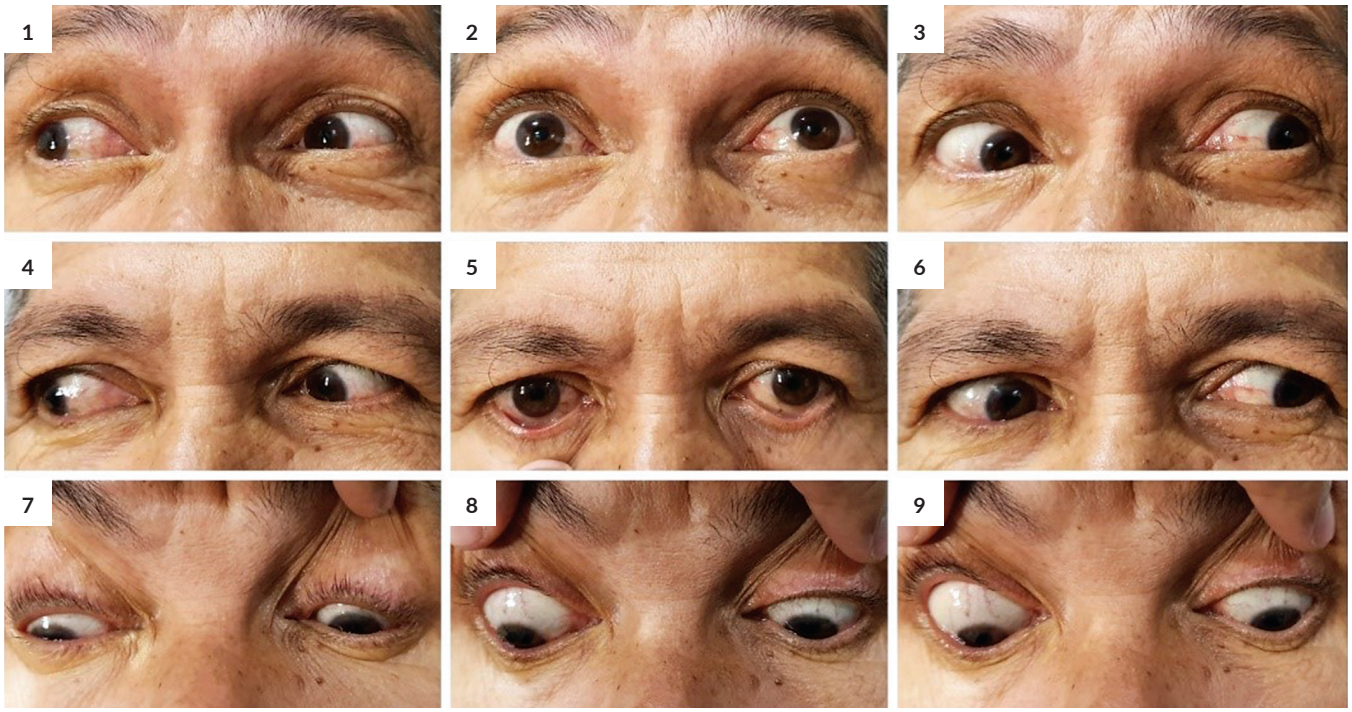

Figure 1. ***Composite Diagram.*** In Frame 5, on primary gaze, there is a slight left hypertropia. Frames 1-3 show conjugate upgaze palsy, the most characteristic feature of dorsal midbrain syndrome. Horizontal (Frames 4 and 6) and downward gazes (Frames 7-9) are preserved.

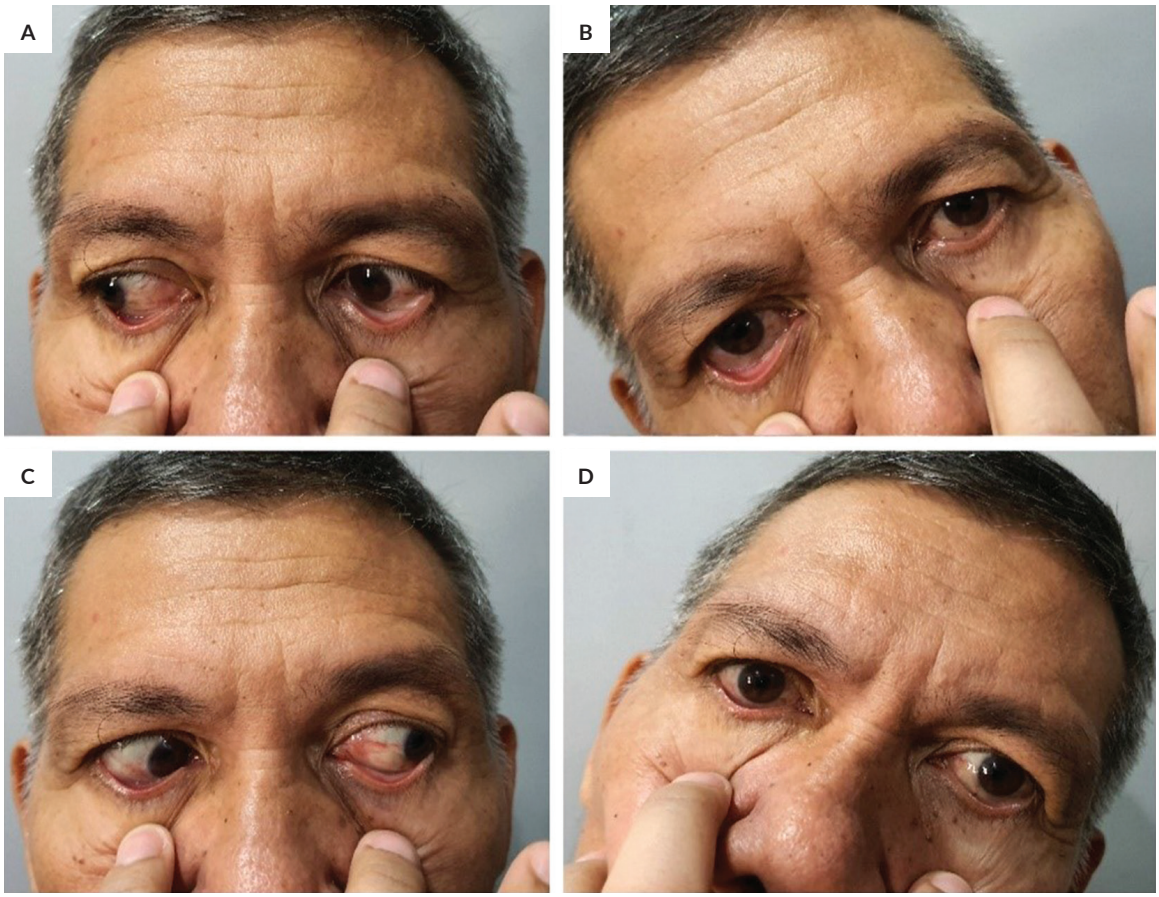

Figure 2. ***Parks-Bielchowsky three-step test.*** There is hypertropia of the left eye on primary gaze. Hypertropia is worse on left-gaze **(A and C)**, indicating a weak depressor *(inferior rectus)* on the left eye or a weak elevator *(inferior oblique)* on the right. Left hypertropia is subsequently worse on left head tilt, indicating a weak incylorotator on the left *(superior oblique)* or a weak excylorotator on the right *(superior oblique)*. No isolated paretic muscle was elicited on this test, pointing to the presence of a skew deviation.

ventricle, and a rightward midline shift (Figure 3). Focal hypodensities likely representing perilesional edema were seen at the tectum, but there was no obvious involvement of the midbrain structures. The rest of the scan was unremarkable save for microvascular white matter ischemic changes and chronic lacunar infarcts at the left internal capsule and left frontal lobe.

DISCUSSION

This patient presented with the classical signs of DMS without clear anatomical involvement of the said structure. Review of available literature has shown only four prior cases of typical DMS from hemorrhages of the thalamic or thalamocapsuloganglionic region.[3-6]

The signs and symptoms of the condition may be brought about by compression of the rostral midbrain and pretectum at the level of the superior colliculus.[1] Currently available literature estimates that up to 65% of cases are secondary to primary midbrain lesions such as infarcts, hemorrhages, and neoplasms, and up to 30% of cases are due to pineal gland tumors pressing on the dorsal midbrain.[7] As mentioned, strokes of the thalamus, located above the midbrain, have rarely been reported as primary causes.[8] DMS from a thalamic infarct in a young person and in a patient with cerebral venous thrombosis have been reported.[3,6] Thalamic hemorrhage as a possible cause has similarly few reported cases, as published by Waga et al. and Lee et al.[7,8]

It has been hypothesized that these thalamic lesions cause DMS through mass effects on the pretectal region and tectum.[1,7] As seen on CT scan, it is likely that the thalamocapsuloganglionic hemorrhage exerted a mass effect on the dorsal midbrain or pretectum which contains the vertical gaze centers, the interstitial nuclei of Cajal (INC) and the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF), thereby causing upgaze palsy.[2] Involvement of the INC, a key structure of the vestibular input pathway, causes skew devation.[9] As the pupillary light reflex fibers synapse at the pretectal nucleus, compression of the region will cause a sluggish pupillary light response.

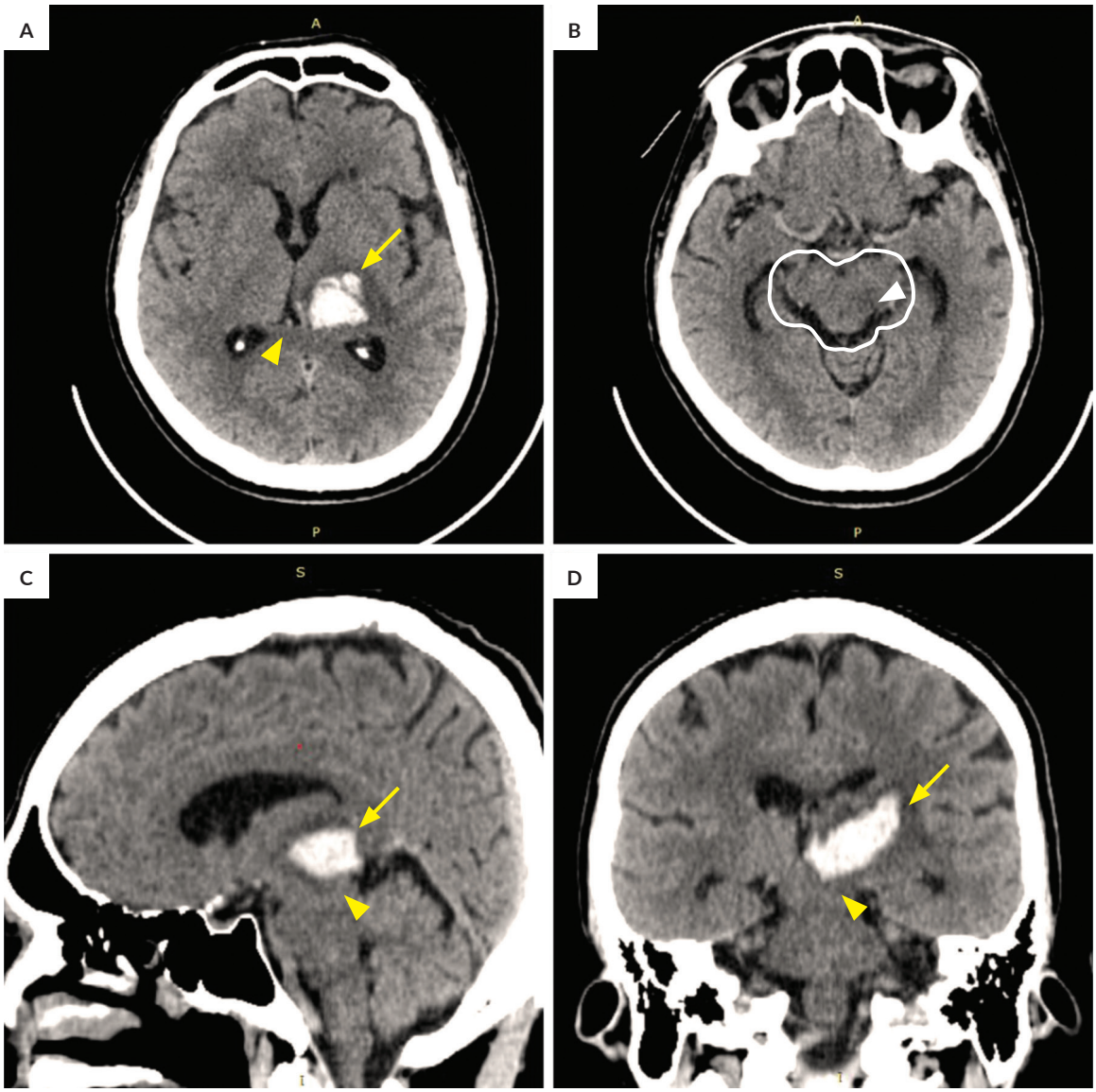

Figure 3. ***Sections from cranial computed tomography scan.*** **(A)** The parenchymal hemorrhage in the left thalamocapsuloganglionic region *(yellow arrow)* was associated with compression of the lateral ventricle and a rightward midline shift *(yellow arrowhead)*. **(B)** Beneath the hemorrhage, focal hypodensities *(white arrowhead)* indicating perilesional edema are seen at the level of the tectum, but there is no obvious involvement of the midbrain *(encircled white, showing normal anatomy appreciated in the classic 'Mickey Mouse' appearance of the midbrain)*. **(C and D)** Sagittal and coronal cuts similarly reveal the location of the hemorrhage *(yellow arrow)* just above the midbrain *(yellow arrowhead)*.

Near reflex fibers are believed to be located ventrally, and are thus spared by a lesion compressing the dorsal midbrain.[6]

Other than direct compression, the affectation of shared vascular supply among dorsal mesencephalic structures and disruption of supranuclear inputs by perilesional edema have also been implicated in the pathophysiology of DMS in cases without direct midbrain involvement.[5,10] Both mechanisms are possibly involved in this case. Supranuclear fibers that control vertical gaze decussate through the posterior commissure as they pass to the rostral interstitial nucleus of the medial longitudinal fasciculus.[1] These fibers have an inhibitory effect on the third nerve nucleus; loss of this inhibition causes sustained discharge from the medial rectus and other extraocular muscles, leading to globe convergence-retraction.[11] These tracts of supranuclear fibers also contain inhibitory fibers to the levator muscle. Damage to these inhibitory fibers can hence cause lid retraction in the primary position (Collier sign), as observed in this patient.

The prognosis of DMS is dependent on the etiology, with literature reporting complete reversal of signs and

symptoms in cases with a manageable primary cause. The case reported by Waga et al. had concomitant hydrocephalus, and the patient displayed complete resolution of symptoms following ventriculoperitoneal shunting.[5] Wang et al. reported complete resolution of symptoms in a patient with DMS from thalamic infarct who was started relatively promptly on aspirin and statins 3-days after the onset of symptoms.[3] Given the scarcity of other similar cases, however, a definite prognostic outlook is still difficult to determine especially in longer-standing cases. Lee et al. had previously reported that complete symptomatic resolution in DMS is unlikely if the syndrome has been present for more than six months.[6] Our patient came in at four months post stroke, and although he reported subjective improvement in his symptoms, he still presented with the classical signs of DMS. Unfortunately, this patient was unable to return to the clinic for scheduled follow-up consult, precluding an adequate reassessment. Ideally, prism glasses could have been prescribed to alleviate diplopia. The patient was able to seek consult via phone call 12 months from his last hospital visit, at which time he reported some additional improvement in symptoms, described as rare, non-bothersome episodes of diplopia. The patient was content knowing that he would have no other gross neurological deficits secondary to stroke. Urgent follow up was not deemed necessary nor was desired by the patient. He was advised and happy to continue observing his symptoms for improvement.

CONCLUSION

This is a case of DMS secondary to a left thalamocapsuloganglionic hemorrhage. The patient's signs and symptoms may ultimately be attributable to mass effects of the primary lesion on the dorsal midbrain, affectation of shared vascular supply among dorsal mesencephalic structures, or compromise of supranuclear inputs to the dorsal midbrain. In any case, this provides further evidence that DMS may result from lesions without obvious involvement of the said region. This case highlights the importance of a complete neurologic examination to elicit possibly subtle signs of a syndromic condition, and the importance of correlating one's clinical findings with a thorough analysis of available diagnostic test results. At the same time, this case provides further evidence that DMS may occur from lesions without obvious midbrain involvement. Thus, it is exemplified as well that the absence of diagnostic confirmation by imaging should not preclude the correct diagnosis by clinical assessment of history and physical examination. The reported persistence, albeit with improvement, of symptoms 16 months post-stroke offers insight into the prognosis of DMS from conservatively managed vascular insults outside the dorsal midbrain itself.

Statement of Authorship

Both authors contributed in the conceptualization of work, acquisition and analysis of data, drafting and revising of manuscript, and final approval of the version to be published.

Author Disclosure

Both authors declared no conflicts of interest.

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