Disappearance of the Hummingbird Sign after Shunt Surgery in a Case of Idiopathic Normal Pressure Hydrocephalus

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Abstract

A 79-year-old man presented with a slowly progressive gait disturbance. Brain MRI demonstrated ventriculomegaly and the hummingbird sign. A lumbar puncture showed no abnormalities of the cerebrospinal fluid. The improvement of the gait disturbance after the ventriculoperitoneal shunt led to a diagnosis of idiopathic normal pressure hydrocephalus. Interestingly, postoperative brain MRI demonstrated the disappearance of not only ventriculomegaly, but also the hummingbird sign. The disappearance of the hummingbird sign suggests that an increase in the cerebrospinal fluid in the lateral and third ventricles could cause the compression of the superior surface of the midbrain tegmentum, which manifests as the hummingbird sign.

Key words: hummingbird sign, idiopathic normal pressure hydrocephalus, progressive supranuclear palsy, rostral midbrain tegmentum

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Introduction

The hummingbird sign refers to a concave aspect of the superior surface of the midbrain tegmentum on midsagittal brain MRI, which is considered to reflect a loss of neurons and fibers in the rostral midbrain tegmentum in cases of progressive supranuclear palsy (PSP) (1-4). Although one study demonstrated the hummingbird sign in cases of possible idiopathic normal pressure hydrocephalus (iNPH) (5), this study did not include definite iNPH cases. Therefore, it remains unclear whether the hummingbird sign is associated with iNPH, and whether it disappears after shunt surgery. We herein report a patient with definite iNPH in whom the hummingbird sign disappeared after shunt surgery.

Case Report

A 79-year-old man consulted the Department of Neurology of our hospital for a 3-year history of slowly progressive gait disturbance. His past medical history included hypertension. On examination, there were no complaints of falls, cognitive impairment, or urinary dysfunction. The patient’s gait was slow and small-stepped, but not wide-based. Gait instability was evident during turning. There was restriction of the upward and downward gaze and mild muscle rigidity around the bilateral elbows. Resting or postural tremors were absent. Brain MRI demonstrated dilatation of the lateral and third ventricles and cerebral aqueduct, thinning of the corpus callosum, and the hummingbird sign (Figure a-e). Evans’ index was 0.32, indicating the presence of hydrocephalus. There was no apparent enlargement of the Sylvian fissure or narrowing of the subarachnoid spaces in the high convexity or medial surface. The callosal angle was steep (less than 90°) on the coronal section through the posterior commissure (Figure e). [¹¹C]FP-CIT single photon emission computed tomography (SPECT) demonstrated that dopamine transporter binding was largely preserved in the putamen (Figure f).

The result of the 3-meter timed up and go test was 71...
seconds [the normative time of subjects aged 76-80 years is approximately 10 seconds (6)]. The Mini Mental State Examination (MMSE) score was 19. A lumbar puncture revealed an opening pressure of 10.5 cm H2O and yielded clear watery fluid with a cell count: 1/μL, total protein: 38 mg/dL, and glucose: 75 mg/dL. After drainage of 30 mL of the cerebrospinal fluid (CSF), the result of the 3-meter timed up and go test decreased to 40 seconds, and the MMSE score increased to 22. Because the CSF tap test ameliorated the gait disturbance, ventriculoperitoneal shunting was performed according to a diagnosis of probable iNPH (7). Thereafter, the gait disturbance improved, and a diagnosis of definite iNPH was made (7). On the other hand, a neurological examination 3 months after the shunt surgery demonstrated that the vertical gaze limitation and muscle rigidity remained unchanged. Brain MRI 3 months after the shunt surgery showed not only the reduction of the lateral and third ventricles and cerebral aqueduct, but also an increase in the width of the corpus callosum and disappearance of the hummingbird sign (Figure g). Evans’ index decreased to 0.27.

**Discussion**

Because the vertical gaze limitation and muscle rigidity remained unchanged after the shunt surgery in our patient, the possibility that a PSP pathology coexisted with iNPH could not be completely excluded. Nevertheless, the disappearance of the hummingbird sign after the shunt surgery cannot be explained by a PSP pathology characterized by a loss of neurons and fibers, and rather suggests that an increase in the CSF in the lateral and third ventricles could cause the compression of the superior surface of the midbrain tegmentum, which manifests as the hummingbird sign. This hypothesis is also supported by the fact that shunt surgery of iNPH increases the anteroposterior and left-to-right diameters of the midbrain (8). Similarly to the midbrain, an increase in the CSF in the lateral and third ventricles was thought to have caused the thinning of the corpus callosum in our case.

According to the findings of the previous (5) and our studies, clinicians should remember the fact that the hummingbird sign can be observed not only in patients with PSP, but also in association with iNPH. Further studies involving a greater accumulation of cases of definite iNPH are necessary to clarify how often the hummingbird sign is observed in patients with iNPH.

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References