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Neuroimaging Highlight

"Hummingbird Sign" Associated with Obstructive Hydrocephalus Due to Aqueductal Web

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A 72-year-old man presented with frequent falls for one year, with worsening balance and gait for the past two months. His family noted urinary incontinence with lack of appropriate social concern and cognitive decline for a month. At presentation, he had an apractic gait with impaired orientation, attention and short-term memory. There was no parkinsonism or ophthalmoplegia. CT scans demonstrated progressive increase in size of the lateral and third ventricles over one year and an MRI was obtained (Figure 1). There was gross enlargement of the lateral ventricles and third ventricle, with a normal fourth ventricle and only minimal periventricular transependymal cerebrospinal fluid (CSF) seepage. CSF flow studies demonstrated absent flow across the aqueduct of Sylvius, and sagittal three-dimensional constructive interference in steady state (CISS) sequences showed aqueductal obstruction due to an aqueductal web. Interestingly, on review of sagittal T1- and T2-weighted images, deformation of the midbrain resembling the "hummingbird sign" described in progressive supranuclear palsy (PSP) was evident.

Aqueductal webs are a rare cause of obstructive hydrocephalus, usually causing symptoms due to raised intracranial pressure in children but may present as adult-onset aqueductal stenosis. The translucent membranous "web" is a septum of fibrillary neuroglia with clumps of ependymal cells across the aqueduct[1] The good image contrast between CSF and brain parenchyma on ultrafast pulse sequences such as CISS and the related fast imaging employing steady-state acquisition facilitates detection of these small intraventricular lesions.[2]

The hummingbird (or "penguin silhouette") sign has been proposed as a specific and sensitive sign of PSP, attributed to atrophy of the midbrain tegmentum.[3,4] The atrophic rostral midbrain is said to resemble the bill and crown of a hummingbird.[4] This subjective sign is associated with a concave-upwards appearance of the floor of the third ventricle and reduced midbrain area relative to the pons and discriminates PSP from healthy controls and patients with other parkinsonian syndromes.[3,4]

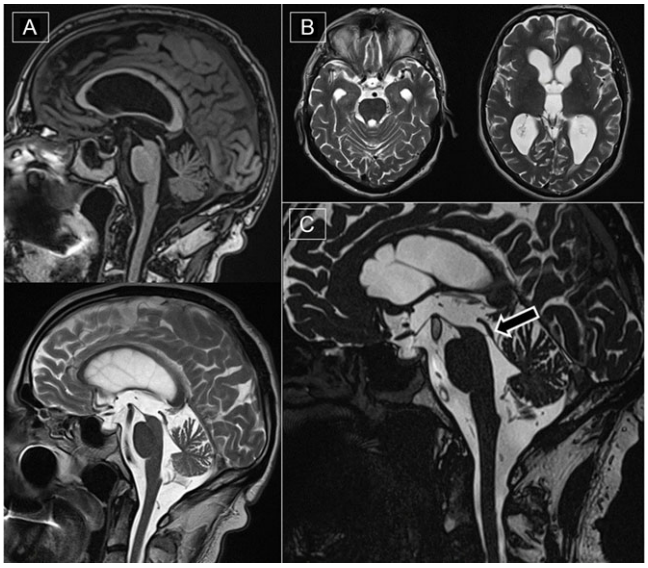

Figure 1: (A) midsagittal T1- and T2-weighted images demonstrating enlargement of the lateral and third ventricles with upward bowing of the corpus callosum and a "hummingbird sign" in the rostral brainstem. (B) Axial T2-weighted images confirm ventriculomegaly with minimal periventricular high signal. The fourth ventricle is normal in calibre. (C) Midsagittal constructive interference in steady state (CISS) sequence confirms the presence of an aqueductal web (arrow) with funnelling of the aqueduct.

However, there is no recognised consensus on cut-off values or mandatory features for the hummingbird sign.[5] In particular, the sign has been reported in patients with non-parkinsonian conditions including idiopathic normal pressure hydrocephalus (NPH) in which the midbrain may visually resemble that of a PSP patient.[5,6] In their studies, Virhammar and Constantinides found that the hummingbird sign was only 43–86% specific for PSP, compared to 95% specificity for a midbrain area <75.3 mm^2.[5,6]

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The lack of transependymal CSF seepage indicates the chronic nature of our patient's hydrocephalus, which led to clinical features usually associated with NPH. To our knowledge, the presence of the hummingbird sign in patients with obstructive hydrocephalus has not been previously reported. This appearance is likely due to pressure effects from the enlarged third ventricle, rather than brainstem atrophy.[5,6]

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