Movement Disorders
CLINICAL PRACTICE

CLINICAL VIGNETTE

Bobble-Head Doll Syndrome in a Child with Suprasellar Arachnoid Cyst

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Bobble-head doll syndrome (BHDS) is a relatively uncommon acquired movement disorder most commonly seen in the first decade of life.[1] It was first reported in children with third ventricular dilatation who exhibited back and forth movement of the head and trunk.[2] These unique rhythmic head bobbing movements at the frequency of 2–3 Hz, which are seen in certain types of dolls, are usually of a "yes–yes" (up and down) type, but are also rarely of a "no–no" (side-to-side) type.[3] The head bobbing disappears during sleep and can be stopped temporarily on command, but worsens during walking or excitement often accompanied with neck, upper limbs, and trunk movements.[4]

Herein, we present an illustrative case of 9-year-old girl with a suprasellar arachnoid cyst that caused BHDS. She underwent endoscopic cyst fenestration, resulting in immediate remission of symptoms.

The patient was admitted to our pediatric neurology clinic with involuntary abnormal head movements. The head shaking started insidiously two years ago and gradually progressed. Constant side-to-side head shaking was most evident with excitement and decreased during talking (Video 1). Her gait was wide-based and clumsy. Side-to-side head bobbing was noted without trunk and limb movements. The patient's height was 126 cm (−1.05 SDS) and weight was 40 kg (1.69 SDS). She also had marked breast enlargement (Tanner 4) and pubic hair (Tanner 4). The rest of the physical examination was normal.

Magnetic resonance imaging (MRI) demonstrated a suprasellar-anterior third ventricular arachnoid cyst and obstructive hydrocephalus. The cystic lesion had measurements of 60 mm in anteroposterior and 45 mm in mediolateral directions. The patient was operated on and cysto-ventriculocisternostomy was performed by endoscopic fenestration. After surgery, most of her head-bobbing disappeared (Video 2). A postoperative follow up MRI showed regression in cyst size and reduction of obstructive hydrocephalus (Fig. 1). At the last visit, her height was 137 cm (−1,8 SDS) and growth velocity was 3,9 cm (−2,6 SDS). She is followed for growth retardation.

Since BHDS was first described by Benton et al. in 1966, there are very few cases reported in literature.[2] The syndrome is usually seen with lesions causing third ventricle dilatation such as suprasellar or third ventricular cysts and aqueductal stenosis.[1,5–7] Other rare pathological abnormalities described in association with this include shunt dysfunctions, trapped fourth ventricle, congenital aqueductal web, Dandy-Walker syndrome, and cerebellar malformations.[1,4,6,8] Endocrinopathies such as precocious puberty, obesity, and growth delay are reported in BHDS.

Although there are a number of theories that explain the underlying mechanism of the syndrome, the exact pathophysiology is still unknown. Whatever the pathogenesis is, the only treatment for this syndrome is surgical. Neurosurgical procedures have favorable clinical results if there is no delay between the onset of bobbing and the surgery.[6,7]

This case report is presented because of its impressive videos and excellent clinical outcome after surgery.

Author Roles

1. Clinical Project: A. Conception, B. Organization, C. Execution; 2. Statistical Analysis: A. Design, B. Execution, C. Review and Critique; 3. Manuscript Preparation: A. Writting the First Draft, B. Review and Critique.

M.S.: 1C, 3A.
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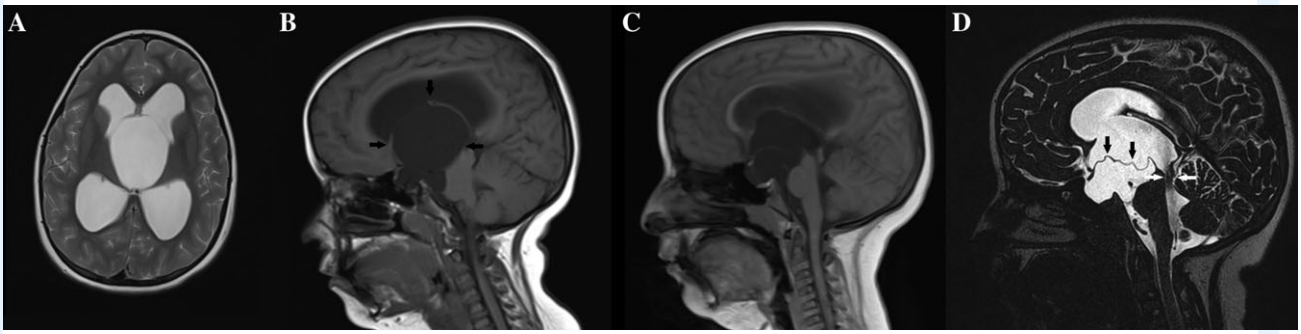


FIG. 1. (A) Preoperative axial T2-weighted magnetic resonance image show the dilatation of third and both lateral ventricles. (B) T1-weighted sagittal image shows that the well marginated, cerebrospinal fluid isointense cystic lesion (black arrows) in suprasellar-prepontine region and obstructive triventricular hydrocephalus with aqueductus sylvii compression. (C) Postoperative fourth month T1-weighted sagittal image shows the cystic lesion and obstructive hydrocephalus have decreased after fenestration. (D) Postoperative first month T2-weighted sagittal image demonstrated the free CSF flow (white arrows) in aqueductus sylvii and decreased cyst size (black arrows).

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Supporting Information

Supporting information may be found in the online version of this article.

Video 1. Nine-year-old girl with no-no type head bobbing that aggravates during walking.

Video 2. After surgery patient's head bobbing almost disappeared.