

Bobble-Head Doll Syndrome

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A 13-year-old girl with aqueductal stenosis (AS) manifesting with “Bobble-Head Doll Syndrome”, a rare movement disorder affecting head and trunk, is reported and the literature reviewed. The abnormal movements improved after shunt surgery.

Key Words: aqueductal stenosis, bobble head doll syndrome, obstructive hydrocephalus

AS is an important cause of hydrocephalus in children. Most often AS is due to congenital lesions i.e. forking of aqueduct, a web or membrane formation or atresia, causing dilatation of third and lateral ventricles. It has characteristics of neurological, psychiatric and endocrine symptoms. However these are overshadowed by the symptoms and signs of raised intracranial pressure.¹⁵ If not recognized and promptly treated in time, it leads to irreversible neurological damage with fatal outcome. Initial presentation as movement disorder involving head, trunk and the body (“Bobble-head doll syndrome”) is very rare and a unique presentation of AS. Only isolated cases are reported in the literature.^{3,11,13,18} Rarity of the condition led to this case report.

Case report

History

This 13-year-old girl presented with poor scholastic performance and involuntary movements characterized by involuntary nodding of head intermixed with side to side movements of 5 years’ duration. In addition, she had involuntary movements of the whole body, at times lurching to either side. However, there was no history of falls during ambulation. In addition she had occasional mild headaches not associated with vomiting, diplopia or visual disturbances. There was no history of seizures. The rest of her history was noncontributory.

Examination

Clinical examination revealed a short stature girl with prominent forehead. The head circumference was 56 cms (95th percentile for her age and sex). She was conscious. Fundus examination showed temporal pallor, more on the

right side. The remainder of the nervous system examination was normal except for the brisk deep tendon reflexes and extensor planter response bilaterally. She had involuntary movements of head, characterized by forward and backward thrusting movements of head accompanied by side to side movements resembling coarse titubation. In addition she had involuntary movements on ambulation consisting of lurching movements of the body at times resembling jumping movements affecting axial musculature. The duration and the speed of these movements varied with time during ambulation and were sensitive to loud sounds and music. There were no movements at rest or during sleep. The movements had a tendency to diminish whenever she was asked to perform a voluntary task. Over the period there was no change in the characteristics of the involuntary movements.

Hospital Course

Routine hematological and biochemical parameters were within normal limits. computerized tomography (CT) scan (**Figure 1**) of the head revealed marked dilatation of lateral and third ventricles with a normal fourth ventricle. A diagnosis of congenital AS was made. The patient subsequently underwent ventriculoperitoneal shunt (VPS) placement. After the procedure the patient had gradual but complete resolution of symptoms.

Discussion

“Bobble-Head Doll Syndrome” was first reported by Benton, et al.,² in 1966 in a child who had hydrocephalus due to third ventricular cysts. The “Bobble-Head Doll Syndrome” is a rare and an interesting movement disorder characterized by continuous or episodic forward and

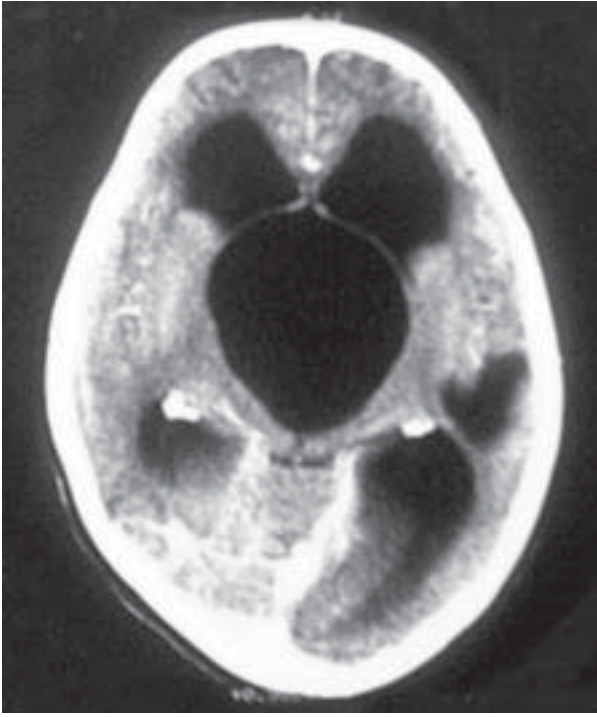


Figure 1. CT scan of head showing obstructive hydrocephalus secondary to aqueductal stenosis.

backward head nodding at times associated with side to side movements of the head of 2-3 Hz frequency. These movements are absent during sleep and transiently disappear or attenuate on volitional activities, often accompanied with truncal tremors. These movements are very sensitive to sensory stimuli.^{2,4} This condition is commonly associated with a lesion in and around the third ventricle. Third ventricular tumors or suprasellar arachnoid cysts are the commonest lesions encountered,^{1, 2, 7, 9, 10, 12, 14, 17} followed by aqueductal stenosis.^{3,11,13, 18} Other lesions cited include communicating hydrocephalus, cyst of the cavum pellucidum and interpositum, trapped fourth ventricle, aqueductal and third ventricular choroid plexus papilloma and developmental cerebellar disorders.^{5, 8, 16} A case is reported following the VPS obstruction.⁶

Though superficially resembling spasmus nutans, the Bobble-head doll movements are different as the former is present in neonatal period and tends to be self-limiting and when pathological it is commonly associated with optic glioma and porencephalic cyst. "Bobble-head doll syndrome" is associated with lesions in and around the third ventricle which are associated with hydrocephalus. It is presumed that this movement disorder is due to the pressure effect of dilated third ventricle which distorts the dorsomedial red nucleus and dentatorubrothalamic pathways.^{5,6,13} In addition, there is compression of medial thalamus, the latter having their somatotopic motor representation of head and neck area.¹³ The pressure over the medial thalamus leads to head and neck tremors, and as the lesion progresses, it gives rise to truncal and appendicular tremors also by affecting thalamic area. Attenuation of tremors on volitional activities and their presence at rest suggest a possible role of basal ganglia in the genesis of this disorder. It is also suggested that the

movement develops as a learned phenomenon as it stops on volitional activities. This probably helps in improving the cerebrospinal fluid (CSF) circulation and is supported by the observation of disappearance of head movements and headache on extreme flexion or extension of neck in a case with Bobble-head doll syndrome.¹⁷

Neuroimaging is essential for evaluation of these patients. Mid-sagittal plane MRI is the best modality for the delineation of CSF pathways and soft tissue. It can also define congenital aqueduct pathology (forking, web membrane or atresia).³ In the present case, neuroimaging pointed toward congenital aqueductal stenosis as the cause. Unlike reported cases, the present case had the onset of symptoms at the age of 11 years. A similar late presentation of Bobble-head doll syndrome in a child with aqueductal obstruction due to a membranous web in its lower part has been reported by Bhattacharya, et al., in 2003.³ As shunting procedures or third ventriculostomy results in complete amelioration of symptoms, the condition should be recognized and treated early. The present case had complete remission of the abnormal movement after shunt surgery.

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This side of neurodiagnosis.....!

This could be a little difficult for the modern day neurosurgeons/neuroradiologists to believe. But an Italian Surgeon had a curious way of diagnosing skull fracture!

In the 15th century, a surgeon by the name of Guido Lanfranc developed a curious way of diagnosing skull fractures that no contemporary physicians were aware of. He could diagnose with a fair degree of accuracy if a person had sustained a skull fracture by playing the head like a violin.

By studying the resonance and instrumentation of violins and other stringed instruments, Dr. Lanfranc realized that the sound made by taut instruments could be altered by the condition of other components.

After some experimentation, he found that he could do the same thing to the human skull by placing a violin string between the person's teeth. When the string was plucked, it would cause the skull to vibrate in a musical note. A clear sound would mean an intact skull, whereas a muffled and dull sound suggested a fracture!

