Bobbling head in a young subject

Kalyan B. Bhattacharyya[1], Sumit Deb[2], S. N. Ghosh[2], S. Mondal[2]

Departments of [1]Neuromedicine and [2]Neurosurgery, Bangur Institute of Neurosciences, Kolkata, West Bengal, India

Abstract

Bobble-head Doll Syndrome is a rare and unique movement disorder found in children. Clinically, it is characterized by a to and fro or side to side movement of the head at the frequency of 2 to 3 Hz. It is mostly associated with cystic lesions around the third ventricle, choroid plexus papilloma, aqueductal stenosis and other rare disorders. An eleven year old child presented in the outpatient department with continuous to and fro movement of the head and declining vision for the last one month. MRI Scan showed a large contrast-enhanced lesion in the region of the third ventricle along with gross hydrocephalus. Ventriculo-peritoneal shunt was inserted and the movements of the head disappeared completely. Bobble-head doll syndrome is a rare condition and therefore this case is presented and the literature reviewed.

Key Words

Bobble-head Doll Syndrome, third ventricular tumour, hydrocephalus, ventriculo-peritoneal shunt

For correspondence:

Prof. Kalyan B. Bhattacharyya, Amrapali Point, Flat 1C, 59F, Bosepukur Road, Kolkata-700 042, West Bengal, India.
E-mail: kalyanbrb@gmail.com

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Case Report

An 11-year-old male subject was referred to our hospital with headache associated with intermittent vomiting for one month, which was followed by sudden appearance of gait disturbances and dimness of vision in both the eyes. There was also sudden development of nodding movement of head for the same duration. He was born of a non-consanguineous marriage, and his birth and developmental history were uneventful. There was no history of unconsciousness. On examination, his higher mental functions were normal. Ophthalmoscopy revealed bilateral papilloedema along with indications of post-papilloedema optic atrophy. Direct and consensual light reflexes were sluggish, and there was evidence of relative afferent pupillary defect bilaterally. He had normal power in all the four limbs, all the reflexes were brisk, and the plantar responses were extensor bilaterally. Examination of the cerebellum revealed difficulty in tandem gait and evidence of dysdiachokinesia; sensory examination was within normal limits. There was no evidence of disorders in ocular motility, nystagmus, tremor, or inappropriate response to sensory stimuli. However, the most remarkable feature was a rhythmic and slow forward and backward as well as side to side continuous bobbing movement of the head at the rate of 3-4 times per second, which was suppressible on command. The movements also decreased when he was asked to carry out simple mathematical exercises and were absent during sleep. There was no obvious evidence of enlargement of the head. Examination of the external genitalia, pubic and axillary hair, and breasts did not suggest any endocrinopathy. In this background, the possibility of bobble-head doll syndrome was considered clinically.

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Investigations

Plain and contrast magnetic resonance imaging study of the brain showed a large-mass lesion involving the pineal region with marked enhancement in post-contrast study. The lesion had measurements of 41 mm, 35 mm, and 41 mm in anteroposterior, mediolateral, and craniocaudal directions, respectively. The tectal plate and the aqueduct of Sylvius were compressed, and the lateral and third ventricles were hugely dilated. Since the lesion enhanced homogenously, germ cell tumor was considered the first possibility in a young subject while others, like pineoblastoma and pineocytoma, were also taken into account [Figures 1 and 2]. Plain radiography of the skull did not reveal any evidence of sutural separation, erosion of the posterior clinoid process, enlargement of sella turcica, or beaten silver appearance; electroencephalography EEG was within normal limits. Visual evoked potential study showed

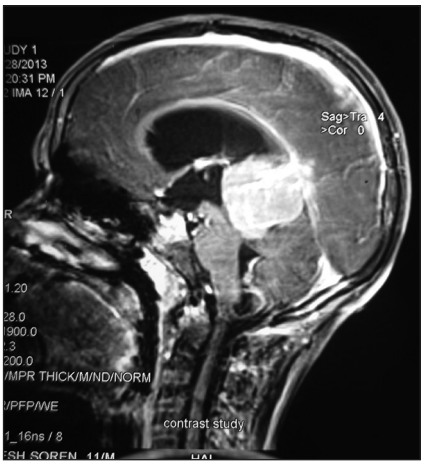

Figure 1: Sagittal T2 contrast image showing an enhancing mass in the posterior third ventricle with hydrocephalus

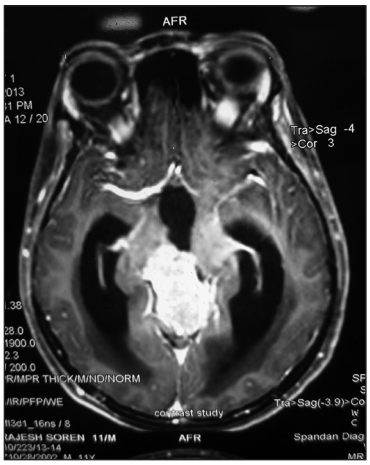

Figure 2: Axial T1 contrast image showing an enhancing mass in the posterior third ventricle with hydrocephalus

evidence of bilateral anterior optic pathway dysfunction in the form of reduction of amplitude, suggesting axonopathy along with some prolongation of the P100 latency, indicating secondary demyelination. Assessment of the serum prolactin, thyroid stimulating, tri- and tetra-iodothyronine, follicular stimulating, and luteinizing hormones were all within normal limits, thus ruling out any possible associated endocrinopathy.

Final Diagnosis

Bobble-head Doll Syndrome

Management

In view of the gross hydrocephalus and the rapidly deteriorating vision, ventriculoperitoneal diversion with medium pressure Chhabra shunt under general anesthesia was planned, followed by removal of the offending tumor. Cerebrospinal fluid collected during the shunt operation was normal in respect to physical, biochemical, and cytological parameters; the study of levels of beta-human chorionic gonadotrphin, alpha-fetoprotein, fetal alkaline phophatase, and lactate dehydrogenase were also within normal limits. Recovery of the patient following surgery was uneventful, and there was no trace of the head bobbing movements thereafter. It has been video recorded for documentation. The dramatic improvement in head nodding led the parents to assume that the patient was completely cured, and they were reluctant to provide consent for further exploration of the offending tumor. The patient was therefore discharged from the hospital.

Discussion

Bobble-head doll syndrome is a rare and unique movement disorder in children and occurs commonly below five years of age.[1] It was first described by Benton *et al*. in 1966,[2] and was soon followed by Nellhaus.[3] It is characterized by continuous or episodic forward and backward head nodding and sometimes, side-to-side movement, usually at the frequency of 2-3 Hz. Neuroimaging studies in most of the situations reveal third ventricular tumor, suprasellar arachnoid cysts, aqueductal stenosis, cystic choroid plexus papilloma of the third ventricle, trapped fourth ventricle, cyst of cavum septum pellucidum, and other lesions in the vicinity of the third ventricle.[1,2,4-7] In recent times, Bhattacharyya *et al*. described a case with a congenital aqueductal web across the third ventricle,[1] and one case has been recorded following the closure of ventriculoperitoneal shunt in meningomyelocoele.[8] The movements disappear during sleep and are eminently controllable under volition.[2] Postural and truncal tremors, nystagmus, and other cerebellar signs are often concurrent features and pyramidal signs in the form of hyperreflexia and extensor plantar response are additional findings. Endocrinopathies of various natures sometimes accompany this condition.[2,4] Extensive reviews of the clinical features, radiological findings, and treatment outcome have been carried out in patients with bobble-head doll syndrome and atypical cases with incomplete clinical presentations have also been described.[1,6-8]

The oldest reported case of bobble-head doll syndrome was 26 years ago.[9] The first case from India was reported by Bhattacharyya *et al*. in 2002.[1] Though Benson *et al*. is universally credited with the first description of the condition,[2] it was Kinnier Wilson who first brought to light a variety of to and fro movement of the head and coined the term *"symptomatic rhythmia"*.[10]

Head nodding is the most important feature of bobble-head doll syndrome and lateral or rotary movements have also been described in addition to the classical movement in six out of 35 cases reported by Mussel *et al*.[4] Suppression of the movements during sleep or under volitional control was a universal feature, and some patients showed extreme sensitivity to sensory stimuli.[2,5] Unconsciousness following drop attacks in congenital aqueductal web or cystic choroid plexus papilloma of the third ventricle has been reported by some workers. This has been attributed to the presence of a semi-permeable aqueductal web interrupting the flow of CSF or due to the mobility of the tumor temporarily blocking its circulation through the ventricular system or the subarachnoid space.[1,11,12] Third ventricular tumor or suprasellar arachnoid cysts were overwhelmingly the most common lesions observed in the imaging studies.[2,4,11,13]

Pathophysiology

The presence of cystic lesions causing swelling in the third ventricle is a common feature in most patients of the bobble-

head doll syndrome, and it is presumed that this dilatation causes pressure on the surrounding structures of the third ventricle, such as the diencephalon. It is also plausible that the back and forth movement of fluid within the cyst causes rhythmic pressure on the diencephalic motor pathways.[2] Swelling of the third ventricle adds pressure upon surrounding structures and thus, their fundamental functions are jeopardized.[3] Weise *et al*. suggested that head nodding was a *'learned phenomenon'* and this view is supported by the fact that the movements are under volitional control.[14] This attests the original contention of Kinnier Wilson that the symptomatic rhythmia is a *'seemingly purposeful movement… often attenuated with pleasurable feeling'*.[9] It is felt that hydrocephalus distorts the red nucleus in the midbrain and the rubrothalamic pathway.[7,8.15] Further studies suggest that dilatation of the third ventricle compresses and thereby distorts the thalamus, which occupies a large paraventricular area. Stimulation studies clearly show that a somatotopic motor representation exists there, the head and the neck being represented most medially and caudally.[6] Therefore, it sounds logical that the third ventricle when compressed, impinges upon the medial thalamus and this probably explains the early development of head and nuchal tremor in this condition.

The case in point presented with some of the classical features of bobble-head doll syndrome and imaging study showed a large mass in the posterior third ventricular area. That ventriculoperitoneal shunt operation ameliorated the condition and vindicated the true nature of the problem. It is also interesting to note that the patient was 11 years old, which is not the usual age for the symptoms to appear.

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