Bobble–head doll syndrome and drop attacks in a child with a cystic choroid plexus papilloma of the third ventricle

Case report

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The bobble–head doll syndrome is a rare movement disorder characterized by 2- to 3-second anteroposterior head bobbing that is reminiscent of bobble–head doll syndrome. This child experienced a sudden onset of drop attacks and then, within several hours, deep coma. The causative lesion was a contrast-enhancing, partially cystic third ventricular mass, which ultimately obstructed the aqueduct, producing profound obstructive hydrocephalus. An emergency ventriculostomy and endoscopic fenestration of the septum pellucidum was performed. Four days later, the tumor was completely resected by a transcavular–transforaminal approach. The lesion was freely mobile within the third ventricle and contained a large cyst within its posterior pole; following drainage of the cyst, the lesion was easily delivered through the foramen of Monro. The histopathological diagnosis was choroid plexus papilloma.

The child’s neurological deficits, head tilt, and head bobbing resolved immediately after operation. To the best of the authors’ knowledge, this represents the first well-documented report of bobble–head doll syndrome and drop attacks secondary to a choroid plexus papilloma. The highly mobile nature of the cystic lesion presumably led to its intermittent impaction within the foramen of Monro and/or proximal aqueduct; this produced the intermittent head tilt and bobble-head symptoms and, ultimately, resulted in acute obstruction of the aqueduct, causing the child’s precipitous neurological decline.

KEY WORDS • choroid plexus • papilloma • drop attack • hydrocephalus • bobble–head doll syndrome • third ventricle • cystic tumor • children
**Preliminary Examination.** The patient showed no evidence of papilledema, nystagmus, visual loss, or focal neurological deficits. A magnetic resonance (MR) image demonstrated pedunculated foci of enhancement within the anterior third ventricle adjacent to the foramen of Monro bilaterally and mild ventriculomegaly with apparent enlargement of the choroid plexus within the left lateral and third ventricles (Fig. 1 left). In retrospect, a cystic-appearing dilation of the posterior third ventricle was visible behind the enhancing areas (Fig. 1). Plans were made for neurosurgical evaluation and repeat imaging in the near future.

Twenty-one days after the preliminary evaluation, the patient was noted by his parents to be having periods of increasing irritability and lethargy; on the morning of the 22nd day, he refused breakfast and had a series of "drop attacks" in which he would fall to the floor for several seconds and then rapidly regain consciousness. Within 3 hours of the initial drop attack, he became comatose and was transported to the emergency room of our institution.

**Examination.** On arrival, he had shallow respirations, disconjugate gaze, dilated unreactive pupils, and decerebrate posturing. He was intubated, hyperventilated, and given mannitol. A computerized tomography (CT) scan was obtained that disclosed severe hydrocephalus as a result of aqueductal obstruction by a lobulated third ventricular mass.

**First Operation.** A left ventriculostomy was performed through a coronal burr hole. The septum pellucidum was fenestrated ventriculoscopically. The left foramen of Monro was completely occluded by a brownish mass that precluded visualization within the third ventricle.

**Postoperative Course.** The patient improved neurologically during the ensuing 3 days. His only residual deficits were disconjugate gaze and mild lethargy. Ventricular drainage ranged from 300 to 500 ml of fluid per day with the drip chamber 15 cm above head level. A repeat MR study disclosed significant enlargement of the third ventricular mass in comparison to the prior study. In addition, the cystic component of the third ventricular lesion had increased substantially in size and now appeared to be tightly impacted within the aqueduct and dorsal third ventricle (Fig. 2 upper left, upper right, and lower left).

**Second Operation.** On the 4th hospital day, the patient underwent a right frontal craniotomy via a transcallosal approach to the lesion. The tumor was impacted within the widely dilated left foramen of Monro with fronds of the lesion extending into the left lateral ventricle. The mass was dissected from around the foramen and mobilized from within the third ventricle. The solid component of the tumor slid freely out of the third ventricle. Immediately thereafter a large cystic portion of tumor spontaneously presented itself at the foramen of Monro; the cyst was substantially larger than the foramen and fully occluded the third ventricular side of the foramen. On puncture of the cyst, the remainder of the tumor was delivered from within the third ventricle without difficulty. The lesion was densely adherent to the medial wall of the left lateral ventricle near the junction of the septal and thalamostriate veins. After mobilization of this attachment, which contained veins draining the tumor and the arterial supply to the lesion from branches of the left anterior choroidal artery, the tumor was removed in one piece.

**Postoperative Course.** The patient awoke with no neurological deficits and had complete resolution of his disconjugate gaze, lethargy, head tilt, and head bobbing. A follow-up MR study disclosed no evidence of residual tumor (Fig. 2 lower right). Permanent cerebrospinal fluid...
Bobble–head doll syndrome and drop attacks

![Fig. 3. Photomicrograph resulting from the histopathological study of a cystic choroid plexus papilloma of the third ventricle neoplasm showing a papillary pattern with uniform cuboidal to columnar cells surrounding a vascularized stroma. Mitotic figures are not visible. H & E, original magnification × 250.]

(CSF) diversion was not required. The histopathological appearance of the tumor (Fig. 3) was typical of a choroid plexus papilloma with a papillary architecture composed of a single layer of cuboidal to columnar epithelium surrounding a vascularized connective tissue stroma; mitoses were not apparent. The patient remains asymptomatic and recurrence free at 15 months follow up.

**Discussion**

Choroid plexus papillomas are benign tumors that are detected most commonly during the first 2 years of life. These lesions typically manifest with macrocephaly and/or gradually progressive symptoms of increased intracranial pressure (ICP) as a result of worsening hydrocephalus. The hydrocephalus stems from a combination of CSF overproduction, intraventricular obstruction, and impaired CSF absorption. Although 10% to 30% of choroid plexus papillomas arise within or extend into the third ventricle, these tumors have not to our knowledge been reported to cause the bobble–head doll syndrome. Choroid plexus papillomas are known to produce tremors. This proposed mechanism is supported by the fact that some patients with the bobble-head phenomenon eventually develop rhythmic tremors of the upper trunk and limbs in addition to the head, implying an origin for the movement abnormality within the extrapyramidal system.

In contrast to hypotheses relating to the above mechanisms, Wiese and colleagues have suggested that the head bobbing may comprise a “learned” behavior, through which the patient is able to reduce cyst size or relieve ventricular dilation by rocking the cyst back and forth, thus intermittently emptying the cyst or allowing CSF to pass around the obstructive lesion. Although seemingly far-fetched, the latter theory is supported by the observation that the bobble-head phenomenon is often positional and may be temporarily suppressed by the patient as it was in our patient. Wiese, et al., discussed a 13-year-old girl, with bobble–head doll syndrome secondary to a suprasellar arachnoid cyst, who reported that her headaches were ameliorated by the bobbing movements. Serial metrizamide CT cisternography studies demonstrated that the arachnoid cyst did indeed decrease in size with head bobbing. The head tilt and forward and backward head bobbing of our patient may conceivably have represented a semivoluntary mechanism of relieving the intraventricular obstruction.

Although the head tilt and bobbing occurred as isolated symptoms in this child, the association of these findings with episodic nystagmus forms a triad referred to as “spasmus nutans.” This entity is generally a benign, self-limited disorder of infancy; however, a percentage of...
patients harbor structural lesions of the retina, optic pathways, or diencephalon. The issue of whether our patient may have had a forme fruste of spasmus nutans without associated nystagmus (sometimes referred to as atypical spasmus nutans) is a semantic one, because neither bobble–head doll syndrome nor spasmus nutans have been previously associated with cystic choroid plexus papillomas of the third ventricle.

In addition, previously described patients with the bobble–head doll syndrome (or, for that matter, spasmus nutans) have not been reported to have had drop attacks or episodes of acute neurological deterioration as a result of sudden ventricular obstruction, presumably because the obstructive lesion in these cases produced gradual rather than acute limitation of CSF flow. Only a handful of nonhemorrhagic intraventricular lesions have been noted to cause precipitous, spontaneous neurological deterioration from acute ventricular obstruction; the most notable examples of this are colloid cyst of the anterior third ventricle and intraventricular cysticercosis. As observed in this child, both of these lesions are capable of abruptly lodging within two areas of the ventricular system in which CSF flow could be effectively halted by a “ball-valve” mechanism, the foramen of Monro, and the aqueduct of Sylvius. Such patients may experience sudden headaches and drop attacks when the head is positioned in such a way that the mass obstructs the ventricle, leading to acute ventricular dilation and a precipitous rise in ICP.

Conclusions

In view of the highly mobile nature of the cystic third ventricular lesion in this child, it is postulated that the tumor, which initially produced head bobbing from intermittent impaction within either the left foramen of Monro or the proximal portion of the cerebral aqueduct, ultimately wedged in and fully occluded the aqueduct, leading to catastrophic neurological deterioration from acute obstructive hydrocephalus. Although the development of a cystic component of a choroid plexus tumor is not unprecedented, the presence of a large, mobile intraventricular cystic component is exceedingly rare, a factor that probably underlies the correspondingly uncommon mode of presentation in this patient.

References


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