Suprasellar Cyst Presenting With Bobble-Head Doll Syndrome

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Abstract

Background: Bobble-head doll syndrome is a rare neurological syndrome presenting with repetitive anteroposterior head movements. It is usually associated with expansile cystic lesions in the third ventricular region.

Case Description: An 8-year-old boy presented with involuntary bobbling head movements. Magnetic resonance imaging of the brain revealed an extensive suprasellar cyst resulting in obstructive hydrocephalus. Endoscopic ventriculo-cysto-cisternostomy resulted in improved clinical outcome.

Conclusions: Endoscopic ventriculo-cysto-cisternostomy is an effective, less-invasive technique in the treatment of suprasellar cysts that results in resolution of the bobbling head movements.

Key words: Arachnoid cyst; Bobble-head doll syndrome; Head bobbing; Suprasellar cyst

Abbreviations and Acronyms

BHDS: Bobble-head doll syndrome
MRI; Magnetic resonance imaging

Introduction

Benton et al.[1] first described bobble-head doll syndrome (BHDS) in children with a third ventricular cyst and obstructive hydrocephalus. The age of onset of BHDS is typically early childhood, although symptoms may appear as early as 1 month or as late as 20 years.[2] The syndrome is caused by lesions causing the third ventricle to dilate, usually due to a suprasellar arachnoid cyst, third ventricular cyst, aqueductal stenosis, or a few other less common conditions.[2,3] Associated neurologic signs and symptoms found on presentation include macrocephaly, ataxia, developmental delay, optic disc pallor or atrophy, hyperreflexia, tremor, endocrinopathy, headache, and vomiting.[2,4]

Case Report

An 8-year-old boy presented with abnormal involuntary bobbling movements of the head which had an insidious onset, progressing over a period of 11 months. The child had no history of headaches, vomiting, or visual disturbances. The child was born via caesarean section due to cephalopelvic disproportion and was normal at birth. His behavioral and intellectual development was appropriate for age. On clinical examination, the child was conscious and coherent. The involuntary bobbling head movement was continuous and more pronounced on walking and absent during sleep (Video 1). He had a broad-based gait with abnormal truncal and knee movements occurring at the same frequency as the head bobbling. His fundus examination and visual acuity were normal with no other focal neurological deficits.

Magnetic resonance imaging (MRI) of the brain showed a thin-walled cystic lesion involving both the suprasellar and the sellar regions. In all sequences, the cystic fluid had the same intensity as the cerebrospinal fluid, with no solid component. The lesion extended into the interpeduncular and prepontine cisterns with splaying of the crus cerebri and mass effect on the pons. The lesion extended superiorly compressing the third ventricle, resulting in obstructive hydrocephalus. The superior extension of the lesion caused splaying of the optic chasm. The normal pituitary bright spot was not visible, possibly secondary to compression by the lesion. Radiologic features were suggestive of a suprasellar arachnoid cyst (Figure 1, Figure 2).

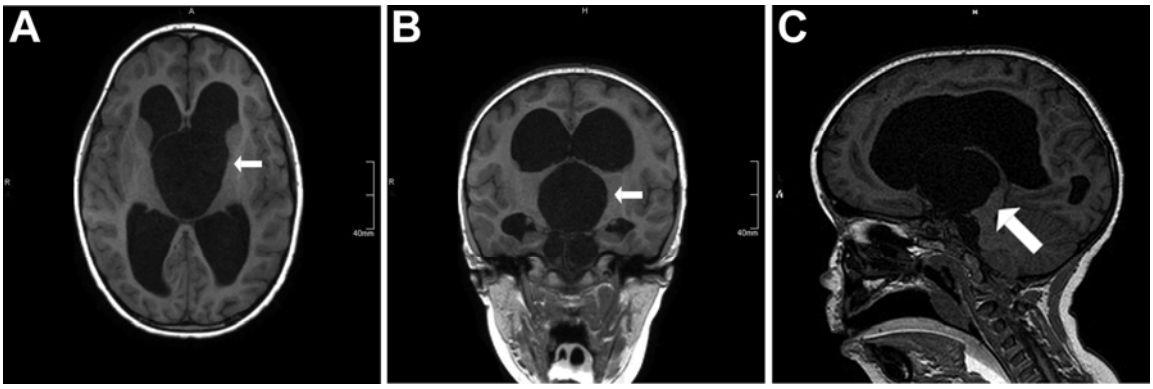

Figure 1. Pre surgical axial (**A**), coronal (**B**), and sagittal (**C**) T1-weighted images demonstrating a large well defined suprasellar cyst (*arrows*) with dilatation of the lateral ventricles.

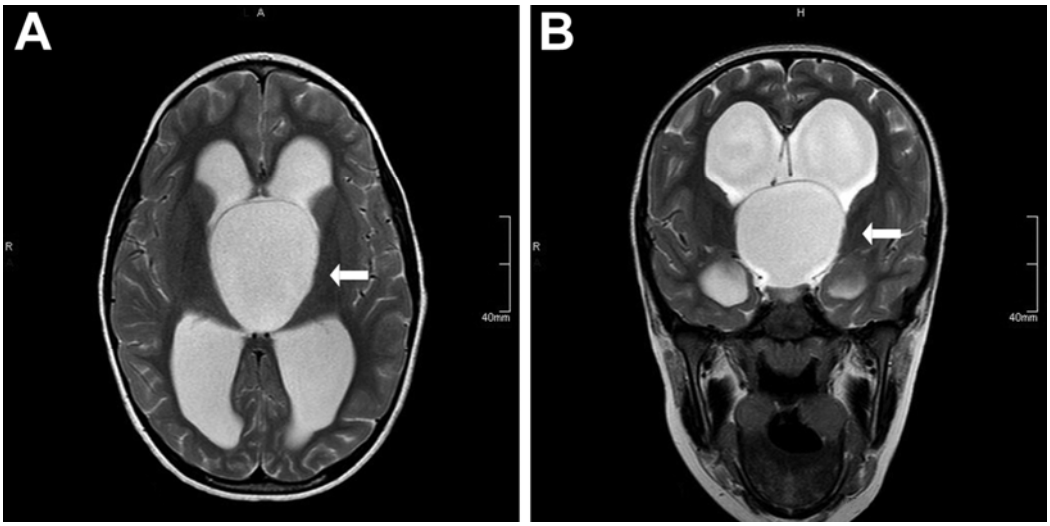

Figure 2. Pre surgical axial (**A**) and coronal (**B**) T2-weighted images demonstrating a large, thin-walled suprasellar cyst with dilatation of the lateral ventricles (*arrows*).

Our intraoperative findings were in agreement with the MRI diagnostics; we noted a grayish cystic suprasellar mass with no solid component to the lesion (Figure 3A). The intracystic fluid was consistent with cerebrospinal fluid. The cyst was not significantly vascular and did not appear to invade ependyma or parenchyma. The cyst wall was biopsied and sent for histology. An endoscopic ventriculo-cysto-cisternostomy (Figure 3B, C) was performed. After surgery, the head bobbling improved significantly, although the movements re-emerged during admission but were significantly less pronounced than pre-procedure (Video 2).

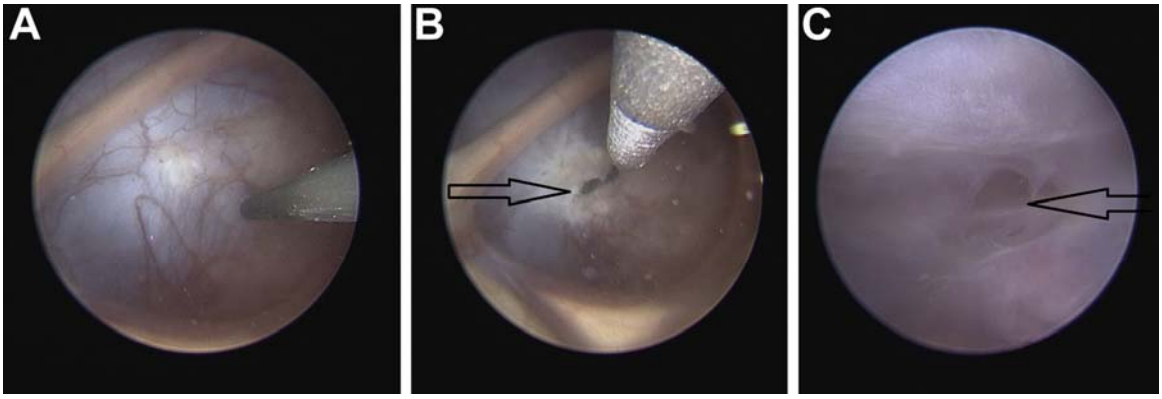

Figure 3. (A) Cyst bulging into the foramen of Monro. (**B**) Ventriculo-cystonostomy (*arrow*). (**C**) Cysto-cisternostomy (*arrow*).

The repeat MRI on day 2 post endoscopic ventriculo-cysto-cisternostomy demonstrated a smaller suprasellar cyst, communicating with the third ventricle and the basal cisterns (Figure 4). The histology of the cyst wall demonstrated high-grade cytological atypia and features were suggestive of a diffuse astrocytoma with cystic change. The histology results conflicted with the radiological and intraoperative findings and we decided to re-biopsy the lesion.

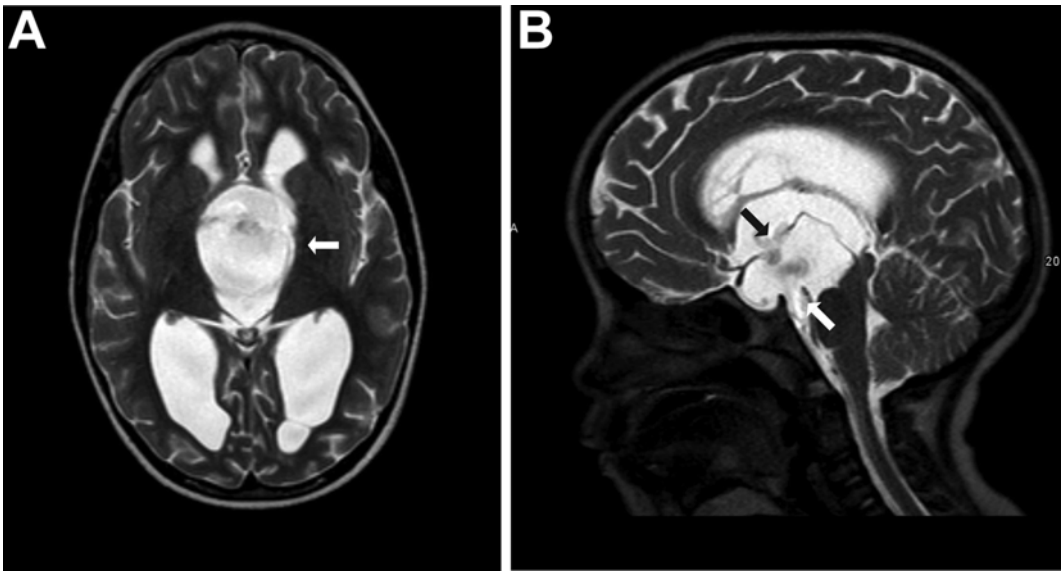

Figure 4. Post endoscopic ventriculo-cysto-cisternostomy; T2-weighted axial (**A**) and sagittal (**B**) images; the suprasellar cyst is smaller and communicates with the third ventricle and the basal cisterns (*arrows*).

At 1 month follow-up, the patient showed barely noticeable head bobbling but still walked with a broad-based gait. There were no new neurological deficits or symptoms of raised intracranial pressure. The patient was doing well at school with no overt neurocognitive impairment. The patient was admitted for re-biopsy of the lesion 4 months later, during school holidays, as requested by his mother. The clinical condition of the patient remained unchanged. Intraoperatively, the cyst wall fenestration and cysto-cisternostomy were still patent. The cyst wall still appeared grayish without evidence of growth. Biopsy of the cyst wall was taken for further histopathologic analysis. The follow-up biopsy was complicated by intraventricular hemorrhage and obstruction of the cisternostomy, however, with resultant hydrocephalus. An external ventricular drain was inserted on day 2 post biopsy. The cisternostomy was endoscopically revised after 3 days; this resolved the hydrocephalus symptoms and the external drain was then removed.

The child developed a right proximal lower limb deep vein thrombosis secondary to a femoral line. He received anticoagulant therapy with warfarin and was being followed up at the neurosurgical outpatient department. The repeat histology of the cyst wall was in keeping with an arachnoid cyst. He was referred back to the peripheral hospital. He completed 3 months of warfarin and a repeat ultrasound of his right lower limb demonstrated resolution of the deep vein thrombosis. Clinically he is doing well, with complete resolution of the head bobbling. He is scheduled for 6-monthly review to follow up his visual acuity, visual field, and pituitary hormone levels.

Discussion

Bobble-head doll syndrome is a movement disorder characterized by continuous or episodic anteroposterior head nodding at times associated with side-to-side movements of the head at a rate of 2 to 3 oscillations per second.[5] This abnormal head movement is reminiscent of a bobble head doll with a weighted head resting on a coiled spring causing the head to bobble with a light tap. The movement increases with anxiety or excitement and decreases or is absent with distraction, when the patient's head is tilted backwards and when the patient is

asleep. The movement can also be voluntarily suppressed for a short period of seconds to minutes in some patients.[2,4]

A recent literature review published in 2018 reviewed 72 cases of BHDS and assessed the associated pathology.[2] Suprasellar cysts accounted for 44% of cases. Third ventricular cysts accounted for 25% and aqueductal obstruction either by an aqueductal web, pineal mass, or tectal tumor accounted for 14% of cases. Less-frequent causes included shunt misplacement or malfunction (6%); other cysts obstructing the third ventricle including septum pellucidum cyst; cavum velum interpositum cyst and a cystic choroid plexus papilloma (4%); masses obstructing the 3rd ventricle, including craniopharyngioma (3%); post fenestration of cysts (3%); and chronic hydrocephalus (1%).

The exact pathophysiology of bobble-head doll movement is unclear. Several theories exist to explain the mechanism of this movement. One of the theories suggests that the cyst compresses the dorsomedial nucleus of the thalamus in the paraventricular region. Compression of the dorsomedial nucleus stimulates the diencephalic extrapyramidal pathways, which include the rubrotegmentospinal and reticulospinal tracts that descend to the motor neurons and innervate the neck muscles, resulting in the bobbling head movements. Another theory suggests that the head bobbling is a learned phenomenon causing intermittent emptying of the cyst, which alleviates the symptoms related to the hydrocephalus.[2,3] Endoscopic ventriculo-cysto-cisternostomy entails endoscopic fenestration of the suprasellar cyst into both the ventricular system and the basal cisterns and is the treatment of the choice for a suprasellar arachnoid cyst with BHDS.[4] Our patient complicated with intraventricular hemorrhage and hydrocephalus. In retrospect, these complications may have been avoided by optimizing the trajectory with neuronavigation and by less aggressive cyst marsupialization, as hemorrhage was noted while working on the caudal end of the cyst.[6]

In this case, the initial histology suggested a diffuse astrocytoma. Surgical removal of gliomas depends on the grading of the tumor, possibility of surgical resection, and patient factors, including age. In this case, if the patient had been diagnosed with a low-grade glioma, the first line of treatment would possibly have been biopsy with cyst decompression followed by adjuvant chemotherapy with or without radiation therapy, in consultation with pediatric oncologist. Although suprasellar gliomas are mostly indolent, tumor progression is observed and so an extensive follow-up plan would have to be put in place. The patient was also referred from a peripheral hospital and many of these patients can be lost to follow-up. It was imperative to re-biopsy the lesion to make a definitive diagnosis as the long-term management of the patient would differ. A patient with a diagnosis of a glioma would require a more complex management strategy with regular ongoing follow-up to monitor for progression.

The histology specimens demonstrated reactive gliosis of the cyst wall associated with reactive atypia. Intracranial gliosis may mimic a low-grade astrocytoma, which can be difficult to distinguish based on histology alone. Hematoxylin and eosin staining may demonstrate cellular atypia in both gliosis and a low-grade astrocytoma (Figure 5A). Immunohistochemical staining using Ki-61, a proliferation marker, may be more useful in differentiating between gliosis and astrocytoma. Reactive astrocytes do not usually demonstrate proliferative activity but may demonstrate a low proliferative rate of 1% to 5% in some non neoplastic conditions, as demonstrated in our case (Figure 5B). Glial fibrillary acidic protein staining was used to confirm the presence of glial cells, which are present in both astrocytomas and gliosis. With gliosis, astrocytes are evenly spaced with thin long glial

processes, whereas with astrocytomas the astrocytes form clusters with shorter, thicker processes (Figure 5C). The low cellularity and the low proliferation index in our case was in keeping with reactive gliosis of the cyst wall.[7,8]

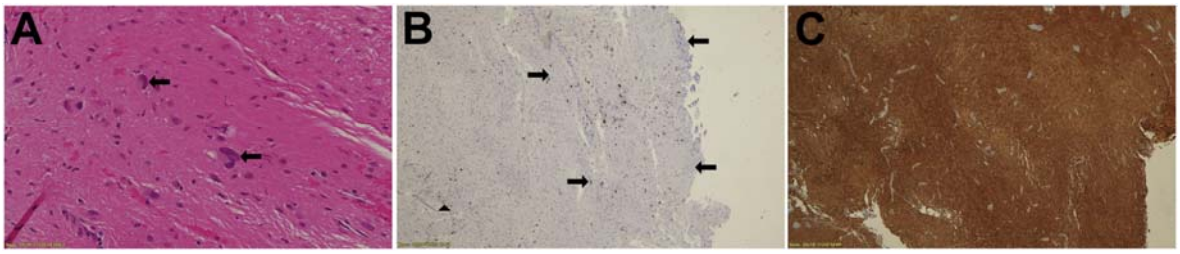

Figure 5. (**A**) Photomicrograph of an hematoxylin and eosin stained section (x40) showing nuclear enlargement and nuclear hyperchromasia in areas of reactive atypia (*arrows*). (**B**) Photomicrograph of sections stained by Ki-67, showing very low proliferative activity of about 1%, of the cyst wall (*region in between the arrows*); surrounding brain tissue (*arrowhead*). (**C**) Photomicrograph of section stained with glial fibrillary acidic protein, showing reactive gliosis.

Conclusions

Suprasellar cysts presenting with bobble-head doll syndrome is a rare occurrence. In this case, the initial histology suggested a diffuse astrocytoma, a tumor, which may have required surgical excision and a more complex management strategy. MRI, intraoperative findings, and the repeat histology confirmed the presence of a benign suprasellar arachnoid cyst that could be managed with an endoscopic ventriculo-cysto-cisternostomy—a safe, less-invasive technique. The patient showed reduced bobbling head movements after the procedure. Histology of brain tumous should always be correlated with radiologic and surgical findings to avoid misdiagnosis of neoplasm.

Acknowledgments

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