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Case Report

Bobble head doll syndrome (BHDS): Case report☆,☆☆

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ABSTRACT

The bobble head doll syndrome is a rare neurological disorder characterized by repetitive and involuntary movement of the head that typically appear in childhood. It is usually associated with the dilatation of the third ventricle and one or more cystic lesions that can be treated surgically. We present the case of a 7-year-old girl with a history of autism, who has experiencing repetitive up and down head movements for 2 years, which were initially thought to be stereotypies. However, 2 months prior to admission, the movements worsened and were accompanied by symptoms of intracranial hypertension. The neurological examination revealed a coordination disorder, specifically a tremor, along with impairment of thermo-algic sensitivity. Ophthalmological examination was unremarkable, but the MRI indicated a colloid cyst of the third ventricle. A minimally invasive neuro-endoscopy procedure was chosen as the treatment of choice for our patient. The bobble head doll syndrome is a complex neurological disorder, and imaging is crucial in the diagnosis and treatment of any movement disorder to enable an early diagnosis and treatment.

Background

Bobble head doll syndrome is a rare neurological syndrome characterized by a repetitive and involuntary head movement disorder that appears in childhood; these movements are anteroposterior up and down (yes-no) and rarely horizontal (no-no) [1–3]. It is usually associated with dilatation of the third ventricle about one or more expansive cystic lesions, and it is rarely associated with posterior cerebral fossa disease; the treatment is surgical [3–5].

Case presentation

We present the case of a 7-year-old girl, who is the second of 3 siblings born to first-degree consanguineous parents, she

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging.

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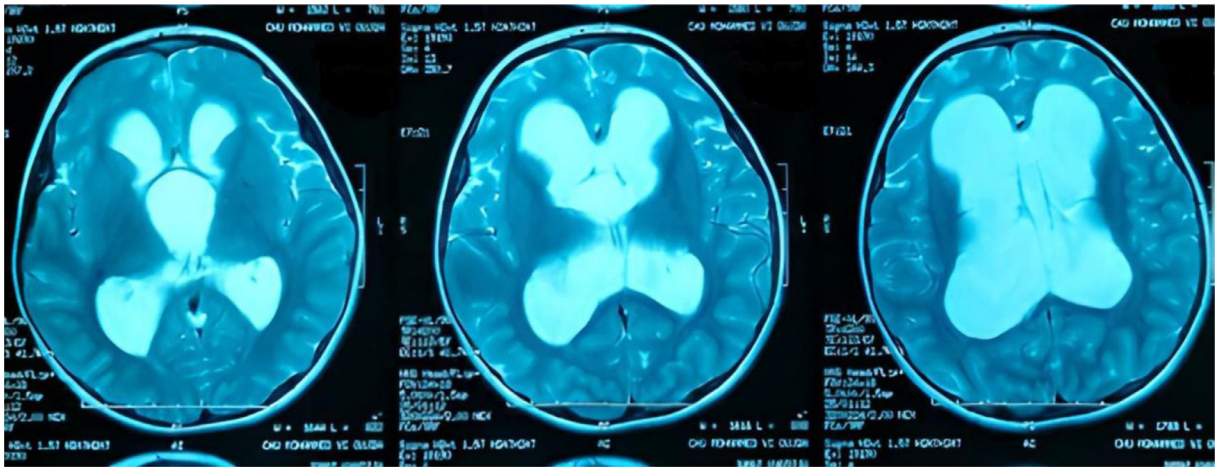

Fig. 1 – Brain MRI axial section T2 shows the cyst in hyper signal with hydrocephalus.

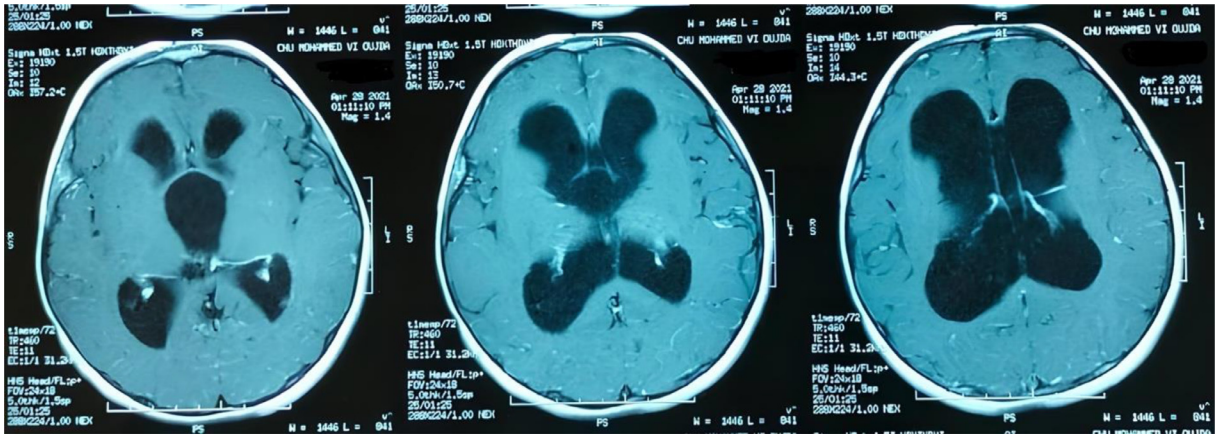

Fig. 2 – Brain MRI axial T1 injected, showing the cyst in hypo signal, not taking the contrast after injection with hydrocephalus.

has a history of autism and began experiencing repetitive head movements to years prior, characterized by an up-and-down of the head from top to bottom (yes-yes) that worsening with annoyance and strong emotions, but disappeared during sleep. The family sought help from a child psychiatrist who diagnosed her with stereotypies related to her autism. However, her symptoms worsened with the progressive onset of headaches, vomiting and a change in behavior, prompting another consultation.

The general examination revealed a conscious patient who was hemodynamically and respiratoryly stable, in good general condition, with normal head circumference, height, and weight, but with signs of early puberty.

The neurological examination showed normal walking and standing, normal global and segmental muscular strength, as well as coordination disorders such as tremor, memory disorders, and altered thermo-algic sensitivity, causing lesions in the patient's upper limbs.

The rest of the clinical examination, including the ophthalmological examination, was unremarkable.

A complete biological workup, including endocrine testing, was normal.

An MRI of the brain revealed a lesion in the third ventricle with a hypo-T1 signal, grossly hyper-T2 signal, not taking the contrast in T1-injected, hypo Flair signal, causing triventricular hydrocephalus upstream, suggestive of a colloid cyst of the third ventricle (Figs. 1, 2 and 3).

A minimally invasive approach using neuro-endoscopy was the procedure of choice for our patient.

Postoperative brain MRI after defenestration showed regression of hydrocephalus (Fig. 4).

The postoperative course was characterized by a clear reduction in the frequency and severity of involuntary head movements.

Discussion

Bobble head doll syndrome is a rare and complex neurological syndrome characterized by repetitive, involuntary head movement that are often up and down with a frequency of 2-3 Hz and rarely horizontal. That movements are stereotypic and rhythmic and they worsen under stress and emotion while

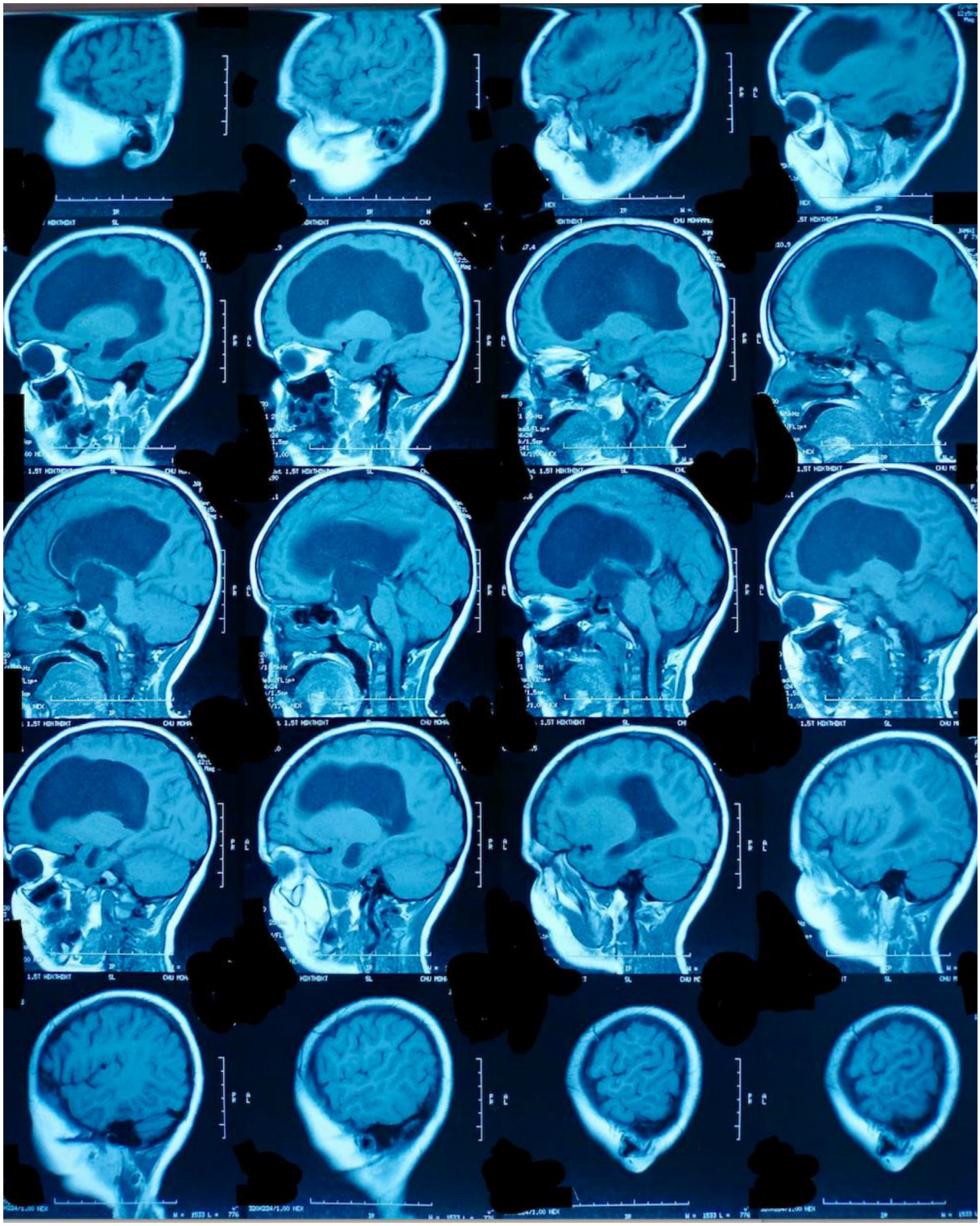

Fig. 3 – Brain MRI sagittal section T1 showing dilatation of the lateral ventricles and the third ventricle with cystic lesion taking the same contrast of the Cerebrospinal fluid.

improving during voluntary movements, tilting the head back, and during sleep. These movements are sensitive to sensory stimuli. It was first described by Benton and colleagues in 1966 [1–3].

In addition to the movement disorder, other symptoms can also be observed in the syndrome such as macrocephaly, ataxia, developmental delay, pallor or atrophy of the optic disk, hyperreflexia, tremor, obesity, endocrine disorders, headache, and vomiting [4,5].

Bobble head doll syndrome is usually associated with third ventricular dilatation, supra-sellar or third ventricular cysts followed by aqueduct stenosis, communicating hydro-

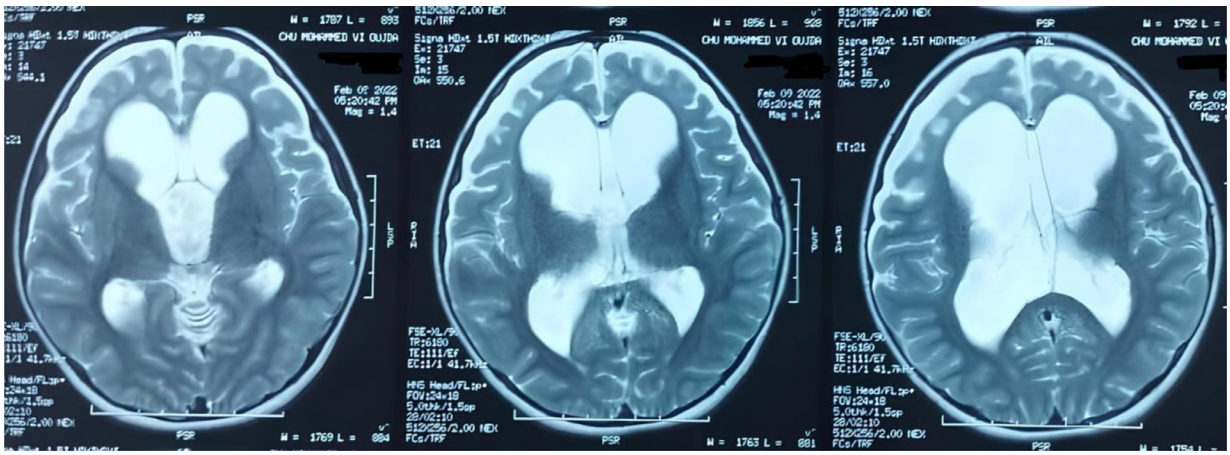

Fig. 4 – Postoperative T2 axial slice brain MRI after defenestration, showing a regression of hydrocephalus.

cephalus, cavum septum pellucid cysts, trapped fourth ventricle, aqueductal and third ventricular choroid plexus papilloma, and developmental cerebellar disorders [5].

The pathophysiological mechanism of bobble head doll syndrome remains unknown and still under discussion and several hypotheses have been proposed, Benton and colleagues postulated that the dilation of the third ventricle caused by the cyst compresses the dorsomedial nucleus of the thalamus in the para-ventricular region and thus disturbs diencephalic structures, Wiese and colleagues suggest that head rocking may be a learned behavior to improve cerebrospinal fluid flow into the third ventricle with a transient decrease in intracranial pressure [6,7].

Neuroimaging is crucial for the diagnosis of bobble head doll syndrome and for postoperative monitoring, Different surgical methods have been described for the treatment of the syndrome associated with a cyst in the third ventricle, including shunting of the cyst, sub frontal approach with ablation of the anterior wall of the cystic lesion, and more recently, the minimally invasive approach by neuro-endoscopy, which involves fenestration of the cyst wall. This approach is currently the best recommended with better results and fewer complications. It has been reported that the delay in diagnosis and therefore in therapeutic management may result in less satisfactory results due to the persistence of the movement disorder and this may be explained by the permanent and irreversible damage to the central structures as a result of the increase in intracranial pressure on the adjacent structures of the third ventricle [8–12].

The most recent review of bobble head doll syndrome was published in 2018, which included reports of 72 cases between 1966 and 2018 [5], Since then, additional cases have been reported between 2018 and 2022, totaling 9 cases [13–21].

Our patient presented with not only the head movement disorder but also other signs and symptoms, including intellectual disability, behavioral disorder, precocious puberty, tremors, and impairment of thermo-algic sensitivity.

This is unique to our case since this sign of impairment of thermo-algic sensitivity has never been reported before and can be attribued the location and volume of the cyst, which compresses both thalami.

During the postoperative follow-up, our patient showed a clear reduction in head movements, tremors, improvement in thermo-algic involvement, and an improvement in behavior.

Finally, it is crucial to emphasize the significance of neuroimaging when facing any abnormal movements. An early diagnosis and treatment can improve the child's psychomotor development and the quality of life of the parents.

Conclusion

The bobble head doll syndrome is a complex neurological disorder. Neuroimaging is essential in the evaluation of any movement disorder and enables early diagnosis and management. The pathophysiology of the syndrome is still not completely understood and is likely multifactorial. Endoscopic ventriculo-cisterno-stomy is the preferred treatment due to its low complications rate and favorable outcomes.

Patient consent

Data collection was made after written parental consent for child. We carried out this case report with respect for patient anonymity and confidentiality of information. (Full written informed consent from parents is available).

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