

Bobble Head Doll Syndrome Revealing A Suprasellar Arachnoid Cyst : Case Report

Siham Bahbouh^{1,2}, Kamel Bouaita³

¹Faculty of Medicine, University of Algiers 1- Algeria

^{1,2}Department of Neurology- Ali Ait Idir Neurosurgical Hospital- Algiers- Algeria

³Department of neurosurgery - Cherchell hospital- Algiers- Algeria

Abstract :

Introduction : Bobble head doll syndrome (BHDS) is a rare neurological syndrome that appears in childhood. It is characterized by abnormal movements of the head, most commonly in an up-and-down motion ("yes yes") and rarely in the horizontal plane ("no no"). We report a case of BHDS revealing a large suprasellar arachnoid cyst.

Observation : A 6-year-old child was brought by his parents to a neurology consultation for abnormal head movements, which most frequently occurred in an upward and downward direction, but disappeared during sleep. These abnormal movements had been progressively worsening over the course of approximately one year. Clinical examination revealed a decrease in visual acuity in both eyes, with papillary hyperemia on fundoscopy and discreet ataxia in walking. Magnetic resonance imaging (MRI) revealed a compressive suprasellar arachnoid cyst with passive biventricular hydrocephalus. Surgical treatment with neuro-endoscopy led to a remarkable regression of abnormal head movements and ataxia in the immediate postoperative period, with complete disappearance in the long term.

Conclusion : Bobble head doll syndrome is a potentially "curable" entity, and diagnosis is provided by cerebral MRI.

Keywords: Bobble head doll syndrome, suprasellar arachnoid cyst, neuro-endoscopy.

Introduction

Bobble head doll syndrome (BHDS) is a rare neurological syndrome that occurs in childhood. It is characterized by abnormal movements of the head, most commonly in an up-and-down motion ("yes yes") and rarely in the horizontal plane ("no no"). We report a case of BHDS revealing a large suprasellar arachnoid cyst.

Case Presentation

A 6-year-old child, with no significant medical history, was brought by his parents for abnormal head movements that started 2 years earlier. The head movements were present during activity and at rest but disappeared during sleep. Over the past year, there had been a progressive increase in abnormal movements, along with diffuse headaches and nausea. Clinical examination revealed a visual acuity of 7/10 in both eyes. Fundoscopy showed bilateral papillary hyperemia. Muscle strength and sensitivity were normal, but there was a mild ataxia in walking. MRI revealed a suprasellar cyst measuring 52 mm in width and 59 mm in anteroposterior axis, with a fluid signal identical to that of cerebrospinal fluid, without

enhancement of its wall after gadolinium injection (figure1,2,3). The cyst occupied the opto-chiasmatic cisterns and exerted a significant mass effect on the V3, causing dilatation of the lateral ventricles, without signs of trans ependymal resorption of cerebrospinal fluid. The cyst displaced and compressed the midbrain posteriorly. There was no intra-sellar extension. The MRI findings were suggestive of a compressive suprasellar arachnoid cyst with passive biventricular hydrocephalus. Biological and endocrine tests were normal. The child underwent surgical fenestration, which led to regression of abnormal head movements and ataxia immediately postoperatively, with complete disappearance in the long term.



Figure 1 : Axial T2-weighted brain MRI showing the hyperintense cyst with hydrocephalus.



Figure 2

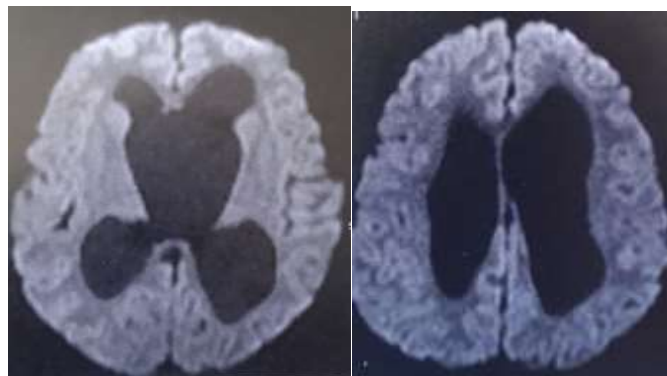


Figure : Brain MRI sagittal T1-weighted (Figure 2) and axial (Figure 3) images showing the dilatation of the lateral ventricles and the cystic lesion with signal intensity similar to cerebrospinal fluid.

Discussion

First described by Benton in 1966, BHDS is a rare and involuntary abnormal head movement. It is most often due to a cyst in the region of the third ventricle but can also be less commonly due to a colloid cyst, a cyst in the cavum septum pellucidum, or a craniopharyngioma. In addition to repetitive and stereotyped head movements, other symptoms reported in BHDS include macrocephaly, pallor or papillary atrophy, ataxia, tremor, endocrine disorders, as well as nausea and vomiting. Reduced visual acuity and/or alterations in the visual field may be due to compression of the nerves and optic chiasm by the cyst wall. The proximity of the cyst to the hypothalamo-hypophyseal area leads to the onset of endocrine disorders. Compression of the diencephalon explains the neurological disorders such as ataxia and BHDS. The pathophysiological mechanism of BHDS is not clear. Benton and colleagues suggested that dilation of the third ventricle caused by the cyst compresses the dorsomedial nucleus of the thalamus in the paraventricular region and disrupts the diencephalic structures. The hypothesis of an extrapyramidal dysfunction, thalamic dysfunction has also been proposed. Finally, Wiese et al. showed that BHDS was a learned behavior to improve cerebrospinal fluid flow in the third ventricle, allowing intermittent drainage with a transient decrease in intracranial pressure. MRI is the imaging modality of choice for diagnosing intracranial arachnoid cysts, showing a clear and regular lesion producing a signal identical to that of cerebrospinal fluid on T1, T2, and diffusion-weighted images, without enhancement after gadolinium injection. The use of neuroendoscopy was the treatment of choice for our patient, providing objective clinical benefit.

Conclusion

BHDS is a potentially "curable" entity. The association of a suprasellar arachnoid cyst with BHDS is extremely rare, and the diagnosis is made by MRI. Treatment with neuroendoscopy is an effective and less invasive technique.

Conflicts of Interest

The authors do not declare any conflicts of interest.

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