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Ependymal Cyst of Third Ventricle Presenting as Intermittent Ataxia in a 2 Year Old Child

Farid Din MD, Muhammad Ubaid Hafeez MD, Khaleeq Ahmed MD and Michael Cohen MD

Department of Neurology, University at Buffalo SUNY, NY, USA

Corresponding author: Farid Din, Department of Neurology, University at Buffalo, SUNY, 100 High Street, E2, Buffalo, NY, 14203, USA. Tel: 7168614969; Fax (716) 859-7833; E-mail: drfarid111@gmail.com**Received:** Dec 16, 2015; **Accepted:** Jan 25, 2016; **Published:** Jan 28, 2016

Abstract

The purpose of this study is to present and document the rare and dynamic behavior of an ependymal cyst. We report 18 months old Caucasian girl who presented with multiple episodes of intermittent ataxia and vomiting with unremarkable neurological examination. Non contrast head CT was consistent with marginally dilated right lateral ventricle. MRI showed 1.7 × 1.5 × 1.1 cm cystic structure at the anterior aspect of the third ventricle at the level of the foramen of Monro. With right frontal endoscopic approach endoscopic ablation of the cyst was achieved. Histologic findings and the immunophenotype was consistent with a diagnosis of ependymal cyst. The patient made an excellent recovery after the procedure.

Keywords: Intermittent ataxia; Ependymal cyst in pediatric population; Non-colloid cyst; Third ventricle; Ball valve mechanism; Obstructive hydrocephalus; Endoscopic ablation

Introduction

Ependymal cysts are rare, neuro-ectoderm derived, benign lesions; mostly diagnosed incidentally on MRI [1]. The most common location is lateral ventricle but they have also been identified in subarachnoid spaces and brain parenchyma [1]. We present a case of ependymal cyst in an 18 months old girl, presented with intermittent ataxia and vomiting. The cyst presented antero-superiorly in the third ventricle; a site common for endoderm derived, colloid cysts (0.5-1% of all brain tumors) but not for ependymal cysts. While ependymal cysts very rarely present in this age group, the unique location and dynamic behavior in our case lead to the intermittent clinical symptoms. Endoscopic cyst ablation resulted in excellent recovery and complete resolution of symptoms.

Case Report

History

An 18 month old Caucasian girl presented with episodic difficulty in walking, intermittent ataxia, broad-based gait and

vomiting. Patient experienced three such episodes, over the course of three weeks. Other than the suggestive ataxia, neurologic examination including fundoscopic examination was unremarkable. Initially, a diagnosis of acute labyrinthitis was considered but excluded following an ENT consultation. Following the third unexplained episode, a non-contrast head CT was ordered which revealed a mildly dilated right lateral ventricle and a third ventricular cyst that was isoattenuating to the CSF (cerebrospinal fluid) (Figure 1). MRI revealed mildly dilated lateral ventricles and an intraventricular cyst 1.7 x 1.5 x 1.1 cm, located anteriorly in the third ventricle, at the level of Foramen of Monro (Figures 2 and 3). The child's symptoms were considered secondary to the cyst occluding the foramen in a ball valve manner, explaining the episodic and paroxysmal nature of the presentation.

Treatment

Patient was admitted to the ICU (intensive care unit) and endoscopic ablation of the cyst was planned. Neurosurgical ablation was performed through a right frontal approach followed by endoscopic third ventriculostomy.

Histology and immunophenotype

The lining epithelium of this cyst consisted of simple cuboidal epithelial cells without goblet cells and no prominent cilia. The epithelial cells showed diffuse positivity for glial fibrillary acidic protein and dot-like positivity for epithelial membrane antigen. Histologic findings and the immunophenotype (negative cytokeratin, positive S100 and variable Glial fibrillary acidic protein immunoreactivity) were consistent with a diagnosis of ependymal cyst.

Post-op Course

Postoperative hospital course was uncomplicated. Complete resolution of symptoms was reported on one month follow-up visit.

Discussion

Third ventricle cysts are almost exclusively colloid and they usually present in 3rd to 5th decade of life [2]. Ependymal cysts hardly ever present in third ventricles, their most common location being lateral ventricles. Third ventricle cysts, although very rare in children, can present as chronic

headaches, papilledema and diplopia[3]. The acute presentation can be in the form of nausea, vomiting, drop attacks and transient loss of consciousness secondary to acute hydrocephalus [4-7].

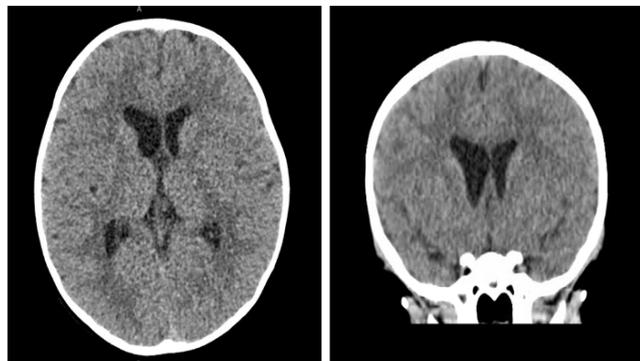


Figure 1 Non Contrast Head CT, Axial (left) and Coronal (right), Slightly dilated right lateral ventricle with evidence of a possible fluid mass obstructing the foramen of Monro

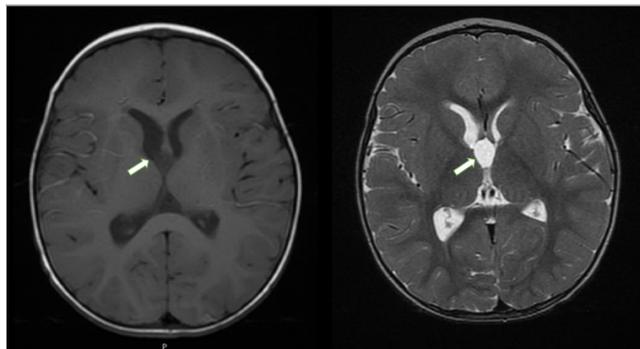


Figure 2 MRI (T1 and T2 Axial views). Hypointense on T1 (left) and Hyperintense on T2 (right), dramatically demonstrating the presence of a mass obstructing the Foramen of Monro

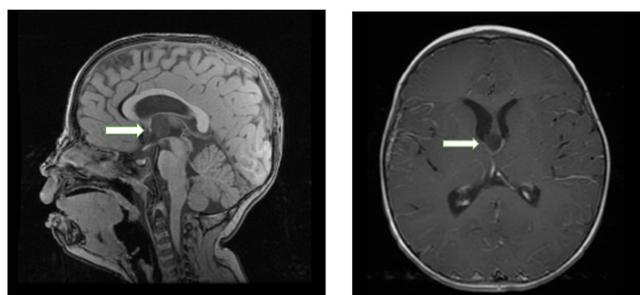


Figure 3 Sagittal T2 FLAIR (left) and Axial T1 Post Contrast (Right). No post contrast enhancement seen on post contrast imaging

Rarely, they have also been described as a sudden cause of death [8-10]. The anterosuperior pendulous attachment in the third ventricle can impart a mobile character to the cyst,

leading to a ball effect resulting in intermittent obstruction of Foramen of Monro. The literature suggests that this mechanism is thought to explain the paroxysmal behavior and clinical symptoms. Of interest, the patient symptoms were not associated with abrupt movements of the head. Symptoms were primarily vomiting and ataxia which goes against Bruns syndrome. The imaging findings effectively rule out metabolic abnormalities, Bruns syndrome, non-specific syncope and bobble-head doll syndrome.

Since third ventricular cysts are usually colloid; they appear hyperdense on CT, hyperintense on T1 and hypointense on T2 weighted images. Signal intensity on neuroimaging depends on the amount of protein content in them [11]. In our case, imaging characteristics did not support a diagnosis of a colloid cyst. The cyst was considered not to be a colloid cyst as it appeared isoattenuating to the CSF on CT scan. On MRI there were abnormal signals identified on all sequences; appearing hypointense on T1, hyperintense on T2 with no diffusion restriction or contrast enhancement.

Asymptomatic cysts should be monitored closely [12]. However, if cysts become symptomatic, surgical treatment should be considered. Among various microsurgical options available for third ventricular colloid cysts, the transcallosal approach is reported to have the most favorable outcome [13,14]. Recent studies show neuro-endoscopic ablation of the cyst to be superior to a microsurgical approach due to low infection risk and smaller number of patients requiring ventriculoperitoneal shunting [12,15]. In our case, endoscopic fenestration of the cyst wall into the subarachnoid space led to quick recovery and complete resolution of symptoms.

Conclusion

Although third ventricular cysts are extremely uncommon in infants and children, they should be considered in the differential diagnosis of intermittent symptoms that are suggestive of raised intracranial pressure. The knowledge of this potentially fatal cause of intermittent ICP in the pediatric population, can hasten the diagnosis. Early endoscopic ablation of the cyst is associated with full recovery and minimal morbidity.

This case in an 18 month old with an ependymal cyst presenting at the level of the foramen of Monro and mimicking signs and symptoms of a colloid cyst may represent the youngest child reported in the literature.

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