

Unusual Cause of Headache in Childhood: Giant Quadrigeminal Cistern Arachnoid Cyst

MF İNCİ, F ÖZKAN, CF DEMİR, ÜE VURDEM

Abstract

Quadrigeminal cistern arachnoid cyst (QAC) is the third most common posterior fossa arachnoid cysts of childhood if middle fossa arachnoid cysts are excluded. These cavities are believed to be congenital and patients usually present with headache, nausea, vomiting, hearing loss, dizziness, behavioural disturbances and psychomotor retardation. QACs are localised between the collicules and cerebellum is usually compressed inferiorly. A 30-month-old boy was referred with vomiting and headache. There was no report of recent associated infections, previous history of any classical childhood viral infection or alterations of psychomotor development. Magnetic resonance imaging showed a large supratentorial arachnoid cyst in the quadrigeminal cistern displacing the brainstem inferiorly and extending anterolaterally into the prepontine cisterns.

Key words

Arachnoid cyst; Quadrigeminal cistern; Magnetic resonance imaging

Introduction

Quadrigeminal cistern arachnoid cysts (QACs) are also known as paracollicular arachnoid cysts, tentorial notch arachnoid cysts, arachnoid cysts of cisterna ambiens, paramesencephalic arachnoid cysts, and parapineal arachnoid cysts. Arachnoid cysts of the posterior fossa represent a rare group of central nervous system lesions. QACs, specifically, are unusual lesions, approximately 100 cases described in the literature.^{1,2}

QACs are the third most common supratentorial arachnoid cysts and occur between the collicular plates and the incisural notch of tentorium.^{3,4} The symptomatology depends on extent and severity of compression on the brain stem, cerebellum and aqueduct.

Classical treatment of such lesions consists of craniectomy and fenestration of the lesion or cystoperitoneal shunting. Neuroendoscopy represents a new effective minimally invasive approach for such lesions.

Herein we report the case of a 30-month-old boy who presented with headache and vomiting due to a large QAC.

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Case Report

History

A 30-month-old boy, presented to our department with one month history of daily headache and vomiting. There was no report of recent associated infections, previous history of any classical childhood viral infection or alterations of psychomotor development. The mother's pregnancy and delivery were uneventful. The examination admission revealed moderate irritable child and macrocrania. Analysis of the cephalic perimeter growth curve showed mild upward deviation since the initial months of life.

Physical Examination

Examination of the eye, ear, nose and throat did not reveal any cause for the headache. During the examination, he was alert, conscious with Glasgow Coma Score of 15 and had no motor deficits. The fundoscopy was normal and there was no loss of any sensory modality in the fifth cranial nerve distribution. The rest of the cranial nerves were all normal. The routine haematology examination was normal.

Imaging Findings

Magnetic resonance imaging (MRI) showed a large supratentorial arachnoid cyst in the quadrigeminal cistern displacing the brainstem inferiorly and extending anterolaterally into the prepontine cisterns, MRI also demonstrated enlargement of the supratentorial ventricular

system secondary to a large QAC compressing the brainstem, cerebellum, aqueduct of sylvius and fourth ventricle (Figure 1). There was hydrocephalus in both lateral ventricles. There was no restricted diffusion on diffusion weighted MRI (Figure 2). The intensity of cyst was same with the cerebrospinal fluid (CSF) intensity in all sequences.

The diagnosis is a QAC with obstructive hydrocephalus and raised intracranial pressure. He was referred to the neurosurgeon for further treatment.

Discussion

Paracollicular arachnoid cyst, cysts posterior to the third ventricle, tentorial notch arachnoid cyst, cyst of cisterna

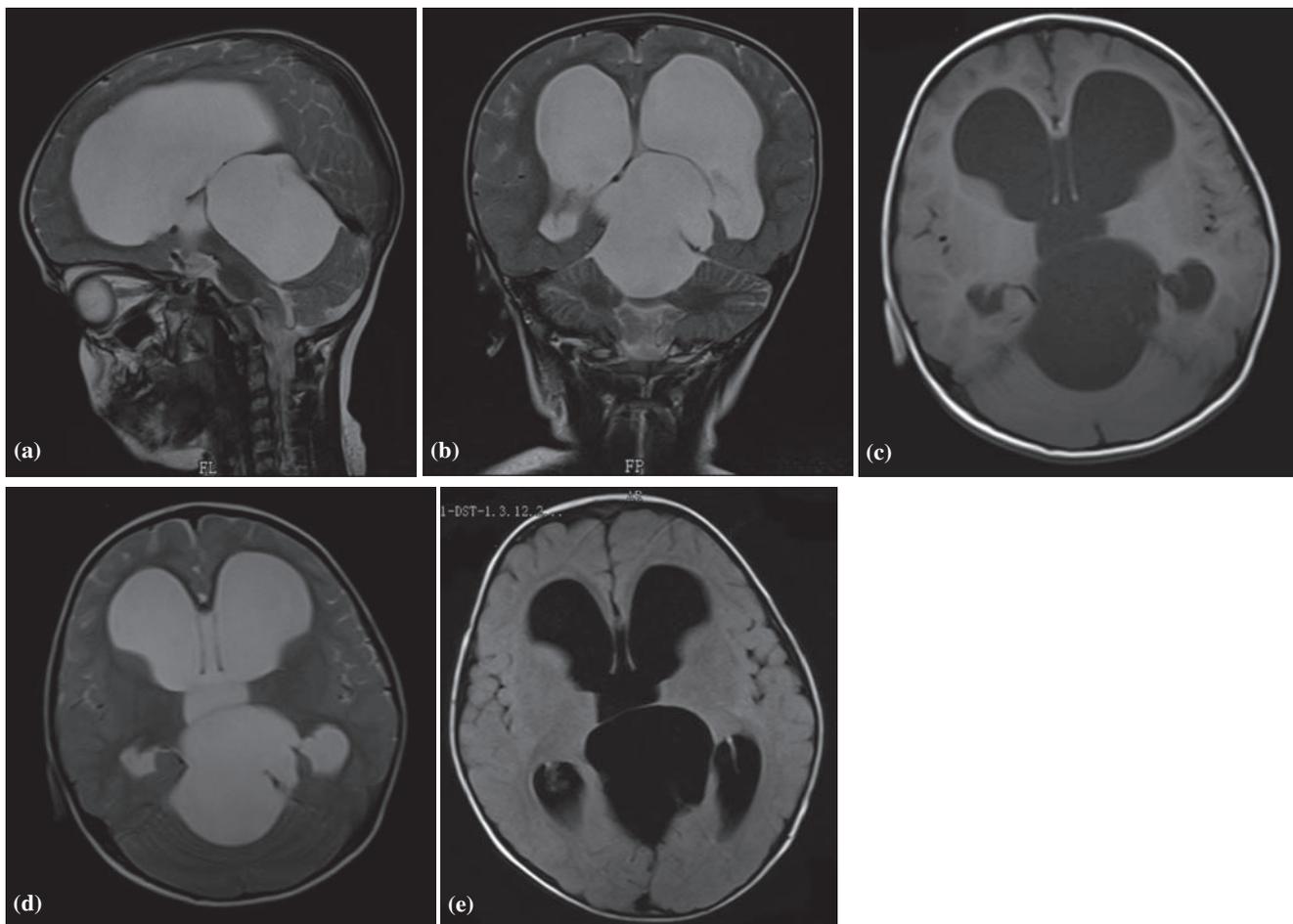


Figure 1 Sagittal (a) and coronal (b) T2 weighted magnetic resonance imaging revealing the cyst abutting the quadrigeminal cistern and causing distortion of the tectum (a), severe compression of the entire brain stem, aqueduct, and cerebellum. Note that the splenium of the corpus callosum is either absent or thinned and the tentorium is elevated (b). Axial T1 (c), T2 (d) weighted and fluid-attenuated inversion recovery (e) images show large quadrigeminal cyst causing hydrocephalus and compressing the brainstem and the fourth ventricle.

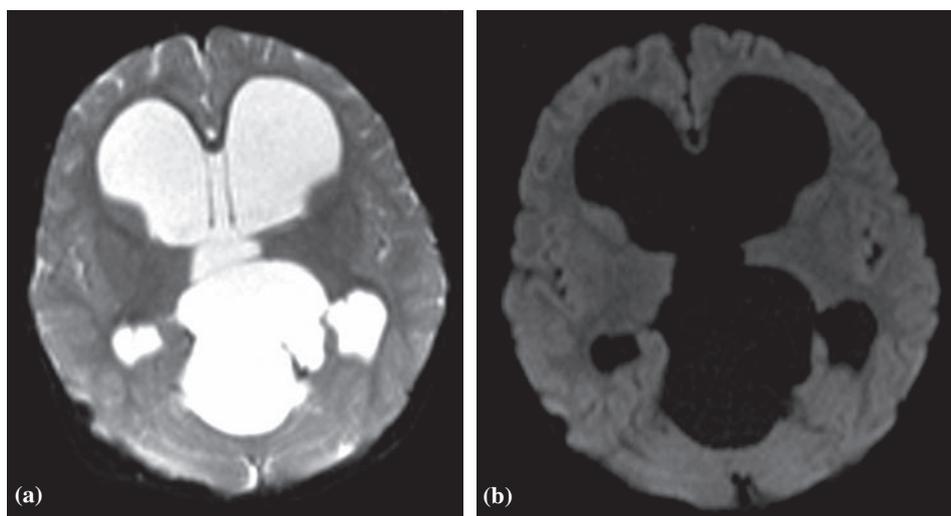


Figure 2 Apparent diffusion coefficient (ADC) mapping (a) and diffusion weighted MRI (b) demonstrating no restricted diffusion on the cyst content.

ambiens, paramesencephalic cysts and parapineal cysts are all synonyms for QACs. Quadrigeminal cistern is a rare location for an arachnoid cyst: there are only about one hundred cases reported in the literature.^{2,5,6} Typical clinical presentation of a QAC includes headache, vomiting, visual complaints, and gait ataxia.⁷ These symptoms are primarily caused by pressure against the tectal plate, which can also compress the aqueduct of sylvius and lead to hydrocephalus. Cerebellar and brainstem compression occasionally occur, as well.⁸

QACs appear as midline, supracerebellar, supratentorial cysts that abut the quadrigeminal cistern on computed tomography (CT) and MRI with no enhancement and presenting the same attenuation values as CSF.³ The enlarging cyst projects downward and backward to lie over the superior surface of the cerebellum. The aqueduct and fourth ventricle are displaced downward and forward and tectal compression occurs.⁵ The third ventricle is distorted and anteriorly displaced. The aqueduct is stretched causing obstructive hydrocephalus.

Generally, the suprapineal recess is elevated and bowed around the cyst, but may extend for a variable distance upward through the tentorial notch, and can be seen on reformatted CT or MRI.¹ The posterior part of the third ventricle is surrounded and displaced anteriorly, although not observed in our case, and the anterior cerebellar vermis is compressed.^{9,10} The aqueduct is stretched and narrowed causing obstructive dilatation of the ventricles.¹¹

The differential diagnoses are lesions which are not suppressed on fluid-attenuated inversion recovery sequence and show restricted diffusion, and other non-neoplastic cysts such as porencephalic cyst, neurenteric cyst, neuroglial cyst. Also pinealoma, aneurysmal dilatation of the vein of Galen, midline meningiomas arising from the posterior tentorial notch and central gliomas invading the quadrigeminal plate are other additional radiological differential diagnoses for this pathology.⁷

QACs may be asymptomatic, or present with symptoms of focal neurological deficit or raised intracranial pressure related to its size and topography. They may present with headache, seizures, macrocephaly or developmental disorders.^{3,12}

Surgical treatment is indicated when the QAC is symptomatic or shows significant mass effect on imaging. Traditionally, QACs have been treated in two ways, either craniotomy and fenestration or cystoperitoneal shunting.⁹ In recent years, neuroendoscopic fenestration of QAC has become more popular as a minimally invasive approach.¹³ Besides, endoscopic cystostomy associated with the third ventriculostomy presents a higher success rate than only endoscopic cystostomy for treatment of QAC.¹⁴

QAC is a rare and treatable pathology in childhood patients. Careful investigation is needed when a child presented with symptoms of intracranial hypertension such as headache, vomiting, visual complaints. MRI is the best diagnostic imaging for QAC.

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