

## CASE REPORT

# Giant Intracranial Arachnoid Cyst Mimicking Benign Paroxysmal Vertigo of Childhood: Case Report with Review of Literature

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## ABSTRACT

Arachnoid cysts constitute 1% of all intracranial space-occupying lesions. These typically produce vague and nonspecific symptoms. However, a subset of these lesions can produce constellation of signs and symptoms indistinguishable from those causing peripheral vertigo. We discuss the presentation of a giant arachnoid cyst which presented with dizziness and mimicked benign paroxysmal vertigo of childhood (BPVC).

**Keywords:** Arachnoid cyst, Vertigo, Subjective visual vertical, Benign paroxysmal vertigo of childhood.

**Source of support:** Nil

**Conflict of interest:** None

## INTRODUCTION

It is very unusual that a child presents with sole complaint of recurrent vertigo. Benign paroxysmal vertigo of childhood (BPVC) is characterized by recurrent attacks of dizziness, with onset between 2 and 8 years of age. The physiopathology of this disorder has been controversial, however majority have pointed out an epidemiological link with migraine.<sup>1</sup>

On the other hand arachnoid cysts are developmental collections of CSF-like fluid covered by arachnoidal epithelium, arising supposedly from congenital malformations. These are mostly located in the middle cranial fossa and present with very vague nonspecific symptoms including vertigo.<sup>2,4</sup>

Herein, we report a clinical case of large arachnoid cyst in the middle cranial fossa which presented with complaints of

only vertigo and mimicked BPVC clinically. It is important to recognize this causal relationship because of the diagnostic and therapeutic implications.

## CASE REPORT

A 5-year-old boy presented to the audiovestibular clinic of our tertiary care teaching hospital with complaints of recurrent brief head rotatory vertigo for 6 months as his only complaint. These were brought about by changes in head positions. During the attacks awareness and speech were not affected. He gave no history of seizures or vomiting. He gave no history of associated headache, hearing loss, tinnitus, and blocked sensation of ears, nasal discharge and blockage. He had no complaints of loss of consciousness or weakness or numbness of any parts of the body. He had no other co-morbidities.

His otoneurological examination was normal except for a sway to left on Romberg's test. Positional tests were negative. Head thrust and head shaking test were negative. Cranial nerve examination was normal. Pure tone audiogram and impedance audiogram were normal. ENG (Electronystagmogram) suggested bilateral hypofunctioning labyrinth (Fig. 1) with right beating nystagmus with head position to left (apogeotropic) (Fig. 2). Video oculography (VOG) demonstrated right beating nystagmus with head position to left. This was fatigable with a brief latency and not direction changing. There was no other positional nystagmus. Static subjective visual vertical and horizontal were within normal limits while the mean dynamic subjective visual vertical and horizontal values were 4.7° and 3.1° respectively.

Magnetic resonance imaging (MRI) was done which revealed a large arachnoid cyst over the left temporoparietal convexity in the middle cranial fossa (Fig. 3) with mass effect evident by gross compression of lateral ventricle (Fig. 4) and significant midline shift (Fig. 5) with minimal hydrocephalus. It was also causing moderate compression of bilateral cerebellopontine angles. However, no effects on the brainstem and cerebellum were evident radiologically. A neurosurgical opinion was sought and parents were counseled about the various treatment options available with probable outcomes and risks involved with each one of

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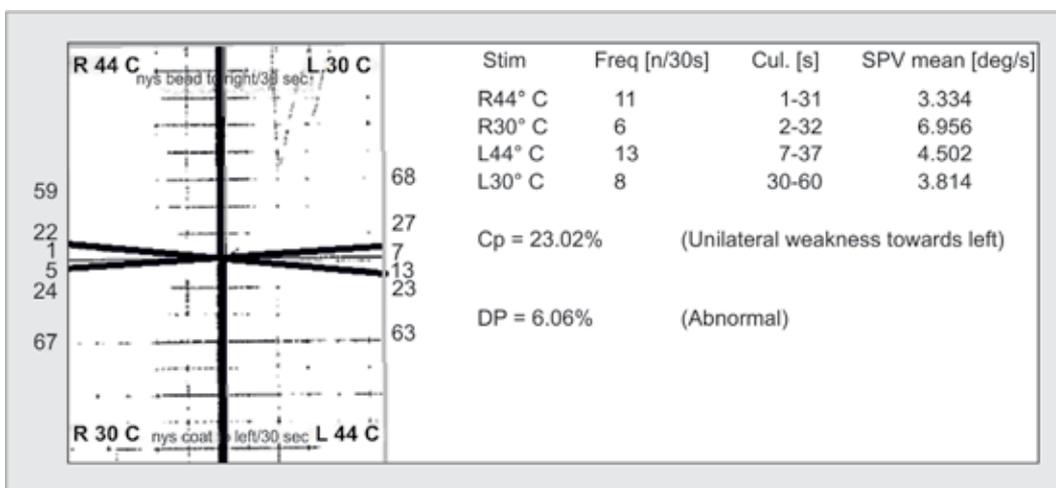


Fig. 1: Electronystagmogram

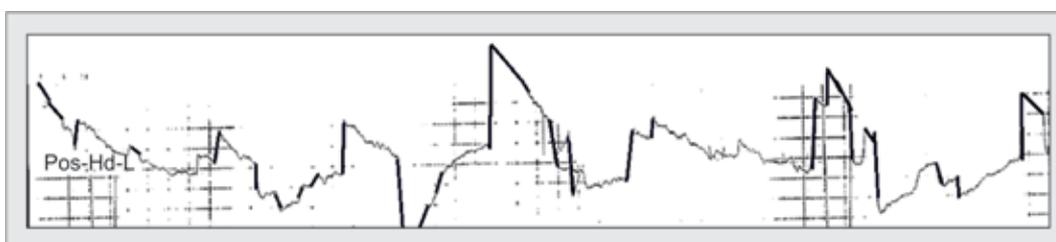


Fig. 2: Right beating nystagmus with head position to left

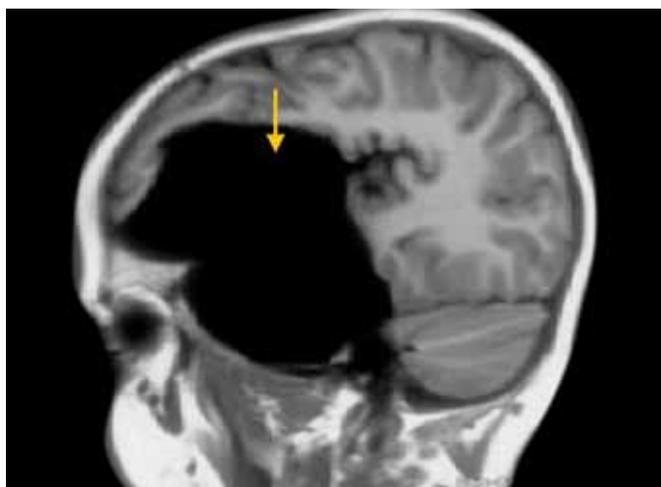


Fig. 3: Arrow showing arachnoid cyst in temporoparietal convexity



Fig. 4: Arrow showing compression of ventricles

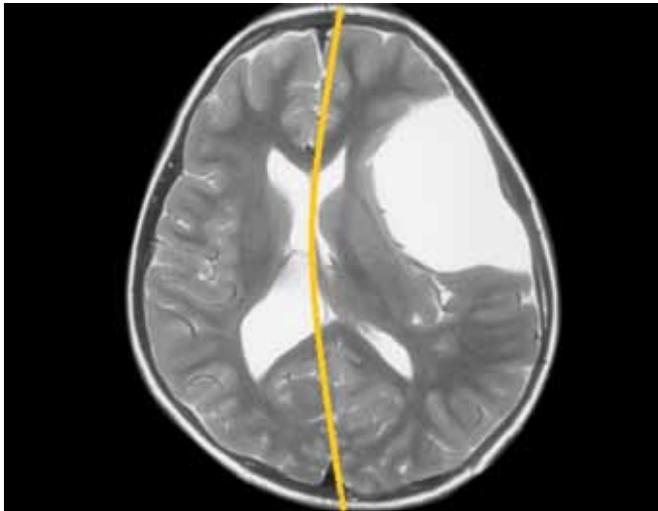
them. The parents decided on conservative management at that point of time with regular follow-up. He was discharged on prolonged vestibular rehabilitation exercises.

**DISCUSSION**

Arachnoid cyst was first described in 1831 by Bright et al as collections of CSF-like fluid encased by arachnoidal epithelium. Most common locations of arachnoid cysts are either in the middle cranial fossa or in the posterior cranial fossa, latter of which is uncommon (5-10%).<sup>3</sup> Arachnoid cysts are usually located in the cerebellopontine angle, cerebellum and the fourth ventricle. They account for

around 1% of intracranial mass lesions. There is no age specific presentation; however male to female preponderance is around 3:1.1. Most are benign in nature and are usually asymptomatic.<sup>3,4</sup> Many are usually more than 5 cm (> 75%) at the time of discovery and can have varied presentation.<sup>4</sup>

Arachnoid cysts in the middle cranial fossa are often diagnosed incidentally, as they are usually asymptomatic and may present with nonspecific symptoms, such as headache, memory disturbance, vomiting, dysphagia, oscillopsia, character change, hearing loss, tinnitus, lower cranial nerve palsies, facial numbness, cerebellar and pyramidal signs, psychomotor retardation and seizures and rarely dizziness.<sup>4</sup>



**Fig. 5:** Midline shift

The posterior fossa arachnoid cysts are often more unexpected and vague. Usually these remain asymptomatic for longer periods and might generate vague and non-specific symptoms which may even mimic multiple sclerosis.<sup>2</sup> Although rarer when compared to middle fossa arachnoid cysts, it has been recognized that cochlear and vestibular deficits may be produced by posterior fossa arachnoid cysts. The 8th cranial nerve dysfunction caused by arachnoid cysts can manifest as hypoacusia, tinnitus and vertigo. Rarely these, arachnoid cyst of cerebellopontine angle, can also manifest as sudden sensorineural hearing loss and even diplopia.<sup>4</sup>

Sometimes location-specific symptoms may be present such as a supratentorial arachnoid cyst mimicking a Ménière's disease attack, frontal arachnoid cysts with depression, left temporal lobe arachnoid cyst with psychosis, left frontotemporal cyst with symptoms of alexithymia. Various other reported associations like cyst on the right sylvian fissure with schizophrenia-like symptoms, cyst on the left middle cranial fossa with auditory hallucinations, migraine-like headaches, and periodic paranoia and left temporal lobe cysts with mood disturbances similar to bipolar disorder have also been rarely reported.<sup>3-5</sup>

Long-standing sensorineural hearing loss and diffuse disequilibrium are associated with arachnoid cysts. Symptoms occur due to expansion or compression of surrounding structures. Mechanisms proposed to explain the above include intracystic hemorrhage, an osmotic gradient leading to passive fluid-diffusion into the cyst, a ball-valve mechanism, or the cyst wall secreting actively.<sup>5</sup>

The etiopathogenesis of arachnoid cysts is still controversial. Congenital theory is the most accepted wherein the cyst apparently results from splitting of the arachnoid membrane cells. Inflammation and trauma have been suggested as acquired causes. Should be before clinical features.

Magnetic resonance imaging is the investigation of choice. Signal intensity is similar to that of CSF. Magnetic resonance imaging also helps in differentiating arachnoid cyst from other cystic lesions. Upon imaging the various differentials to be kept in mind are mega cisterna magna, epidermoid cyst, subdural hygroma/chronic subdural hemorrhage, cystic tumors like pilocytic astrocytoma and hemangioblastoma, non-neoplastic cysts like neurenteric cyst, neuroglial cyst and porencephalic cysts.<sup>6</sup>

Controversy exists regarding the lines of management; surgical treatment vs a watch and wait policy. Various surgical treatment options for arachnoid cysts have been reported. Cyst fenestration, stereotactic puncture, endoscopic cyst fenestration, cysto peritoneal shunt, cyst marsupialization, and complete or partial resection of the cyst wall are few of the surgical options for arachnoid cyst.<sup>7</sup> Surgical intervention usually improves vestibular symptoms however the auditory deficits respond to a lesser extent. However, various studies have also preferred surgical management over the conservational therapy in as high as 75% of the cases. The surgical management has been upfront by mainly excision and marsupialization, stereotactic aspiration, endoscopic fenestration and by cyst peritoneal shunting. Out of the above listed techniques surgery excision and marsupialization of symptomatic cases have provided better results.<sup>8</sup> However, seizure, infection, coma, bleeding, stroke, blindness and even death are few of the listed complications associated with the above said surgical procedures.<sup>2</sup> On the other hand patients with headache have shown remarkable improvement in as high as 86% of the patients. Similar encouraging results are reported in patients with hydrocephalus, gait defects, cognitive disorders and also in patients with seizures.<sup>9</sup>

It has been also well documented in literature that arachnoid cysts should be conservatively managed as far as possible in cases where the patients do not exhibit signs of increased intracranial pressure or focal neurological signs and surgery should be only reserved for only refractory cases and symptomatic patients.<sup>10</sup>

Therefore not all arachnoid cysts require surgical intervention. The benefits and risks of surgical treatment should be discussed with the patient and choice of the management has to be individualistic and justified.<sup>7</sup>

Vertigo in patients with arachnoid cyst may be a consequence of vascular compromise, altered CSF dynamics, and displacement of the VIII nerve, compression of cerebellopontine angle or compression of the endolymphatic sac.<sup>4</sup> Vestibular symptoms by far are more common than cochlear symptoms. Our patient had a dizzy spell, during examination in the left lateral position with right beating nystagmus; this finding and the paroxysms of vertigo in the absence of hearing loss

classifies it as BPVC a condition that is thought to be related to migraine.<sup>11</sup> Dizzy attacks are brought on by change of head position.

Benign paroxysmal vertigo of childhood is classically characterized by recurrent, brief episodes of vertigo that occur without warning and resolve spontaneously after minutes to hours.<sup>11</sup> The age of onset is usually between 2 and 8 years. Vertigo is the initial and primary feature of the attack and is usually not manifested as altered consciousness or headache.<sup>12</sup> However, nausea or other abdominal discomfort may follow. Usually the episodes last for seconds or minutes. Nystagmus may accompany the vertigo, but is often not reported. The neurologic examination is otherwise normal.<sup>11</sup> The above features were present in our case. However in view of atypical positional nystagmus, imaging was done which revealed a large middle fossa arachnoid cyst.

Clinical evaluation and history forms the basis of diagnosing benign paroxysmal vertigo of childhood. Still, intracranial tumors, cervical spinal problems, otological pathology, epilepsy (benign occipital epilepsy), or metabolic disorders should be kept in mind as potential differentials.<sup>11</sup> Treatment of BPVC includes pharmacological and non pharmacological options. Nonpharmacological options include behavioral therapy and vestibular rehabilitation exercises.<sup>11</sup>

## CONCLUSION

Symptomatic arachnoid cysts remain rare in the pediatric age group, which is probably due to their very slow development. Moreover, vertigo as the main presenting symptom is still rarer in children and can mimic BPVC. Children with atypical positional vertigo need to be imaged to rule

out other intracranial pathology. In a majority of patients with arachnoid cyst regular follow-up with conservative management is useful.

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