Cerebellopontine angle arachnoid cyst

Epidemiology

Middle cranial fossa is the most common site of intracranial arachnoid cysts, followed by the cerebellopontine angle (CPA) and suprasellar area.

Gardner et al., in 1960 attributed an embryonal atresia of the fourth ventricle as the cause of "arachnoid cyst" of the cerebellopontine angle.

Clinical features

They usually remain asymptomatic so they are often diagnosed incidentally during radiological evaluation for other reasons.

As these cysts enlarge, they may compress surrounding structures and cause neurological symptoms. Patients may present with vague, nonspecific symptoms such as headache and ataxia.

These cysts also can cause dysfunction of specific cranial nerves, including III, IV, VI (to cause diplopia), V (to induce trigeminal neuralgia), VII (to cause congenital or acquired facial paralysis), VIII (to cause hearing loss, tinnitus, vertigo), X (to result in hoarseness and dysphagia).

Gurkas et al. report a patient with cranial nerve palsies and mirror movements found in upper extremities. They postulated that CPA arachnoid cyst compressing the brain stem and the pyramidal decussation may lead to mirror movements.

Diagnosis

MRI (magnetic resonance imaging) scan techniques have led to CPA arachnoid cysts being more frequently diagnosed and with a higher degree of certainty. The need for further understanding of their natural history as well as for the development of a management rationale has been highlighted with this increased rate of diagnosis.
In the series of Alaani et al. these lesions have a characteristic location in the posterior-inferior aspect of the CPA below the facial and vestibulocochlear nerves. These cysts did not show change in size on repeated MRI scan and the patients' symptoms did not progress over the period of follow up.

**Differential diagnosis**

A cerebellopontine angle lesion could be a vestibular schwannoma, meningioma, epidermoid cyst, or less likely, arachnoid cyst, metastasis, lower cranial nerves schwannoma, lipoma, hemangioma, paraganglioma, or vertebra-basilar dolichoectasia. Primary meningeal melanocytoma is a rare neoplasm, especially when it occurs at the cerebellopontine angle.

MRI is helpful in differentiating arachnoid cysts from those cystic lesions. If a pathologic cause of a retrocochlear disorder is suspected in a patient with a unilateral sensorineural hearing loss and tinnitus, MRI should be performed to evaluate the cerebellopontine angle.

On MRI, arachnoid cysts appear as smooth-surfaced lesions that in all magnetic resonance sequences exhibit a signal characteristic of CSF. In contrast, epidermoid cysts show mixed signals on FLAIR images and high signals on diffusion weighted images. Neurenteric cysts present high signals on T1-weighted images and cystic schwannomas show some foci of contrast enhancement on T1-weighted postcontrast images.

The rising of a neuroglial cyst from the nerve sheath is a finding that brings other possible origins of neuroglial cysts into consideration.
Treatment

The optimal surgical management of arachnoid cysts remains controversial. Although surgery for these entities is controversial, arachnoid cysts can be treated surgically with open craniotomy for cyst removal, fenestration into adjacent arachnoid spaces, shunting of cyst contents, or endoscopic fenestration.

Alaani et al. support a conservative management approach to the majority of these cysts.

The definitive treatment for these arachnoid cysts is a retrosigmoid suboccipital craniotomy and microsurgical resection and fenestration of the cyst walls.

Outcome

The risks of surgery are few, but complications (meningitis, hemiparesis, oculomotor palsy, subdural hematoma, grand mal epilepsy, and death) have been reported.

Olaya et al. report the first case of complete recovery from sensorineural hearing loss and facial weakness following endoscopic fenestration.

Case series

3 pediatric patients with CPA arachnoid cysts (2 with hearing loss and 1 with recurrent headaches) who underwent neurosurgical treatment at the authors' institution.

Four pediatric patients were diagnosed with CPA arachnoid cysts at the International Neuroscience Institute during the period from October 2004 through August 2012, and 3 of these patients underwent surgical treatment. The authors describe the patients' clinical symptoms, the surgical approach, and the results on long-term follow-up. RESULTS One patient (age 14 years) who presented with headache (without hearing deficit) became asymptomatic after surgical treatment. The other 2 patients who underwent surgical treatment both had hearing loss. One of these children (age 9 years) had recent-onset hypacusia and experienced complete recovery immediately after the surgery. The other (age 6 years) had a longer history (2 years) of progressive hearing loss and showed an interruption of the deficit progression and only mild improvement at the follow-up visit.

CPA arachnoid cysts are uncommon in pediatric patients. The indication and timing of the surgical treatment are fundamental, especially when a hearing deficit is present.

2005

Alaani et al. present a series of five adult patients with different clinical presentations attributed to CPA arachnoid cysts. These lesions have a characteristic location in the posterior-inferior aspect of the CPA below the facial and vestibulocochlear nerves. These cysts did not show change in size on repeated MRI scan and the patients' symptoms did not progress over the period of follow up. The findings would support a conservative management approach to the majority of these cysts.

1997

Five patients (three male and two female patients) with a mean age of 5.6 years have been operated on at the Department of Neurosurgery, New York University Medical Center, USA. since 1980 till 1997.
All five arachnoid cysts compressed the cerebellum or brain stem. One patient had associated hydrocephalus. Three patients presented with refractory headaches associated with nausea and vomiting. The remaining two patients presented with cerebellar signs. No patient had an initial cranial neuropathy.

All patients underwent a retrosigmoid suboccipital craniotomy and microsurgical resection and fenestration of the cyst walls. One patient underwent two procedures. A cystoperitoneal shunt was inserted at the first operation. After the shunting procedure, the patient’s condition deteriorated; however, after the microsurgical resection and fenestration, his symptoms improved. With a mean 5.2-year follow-up, there has been no evidence of clinical or radiographic recurrence.

1992

Two cases of arachnoid cysts of the cerebello-pontine angle. The otologic symptoms were unsteadiness, hearing fall and tinnitus. In the first case, the patient who presented a cerebellar syndrome was operated. Afterwards the hearing fell and he developed a transient hydrocephalus. The symptoms disappeared in 9 months. In the second case, the patient was not operated. She was treated medically and supervised. Then the symptoms disappeared too. The authors review the paraclinic exams especially MR, relevant to the diagnosis and discuss the opportunity of a surgical operation.

1984

Ten cases of arachnoid cysts of the ponto-cerebellar angle are presented. In most cases, local arachnoiditis is disclosed during surgery, directing the discussion toward acquired pathology, and perhaps toward local infection. The clinical symptoms are dominated by the cochleo-vestibular deficit, but the involvement of the V and the VII cranial nerves is inconstant and discreet. At the present time, C.T. scan allows a rapid diagnosis but the prognosis must be reserved, in account to the possibility of a local post operatory arachnoiditis.

Case reports

2017

A 4-year-old boy with global developmental delay, esotropia, moderate aortic root dilation, genu valgum, and in-toeing gait. MRI brain for evaluation of neonatal hypotonia revealed a left cerebellopontine angle arachnoid cyst. He referred on newborn hearing screening, and diagnostic auditory brainstem response (ABR) showed left profound retrocochlear hearing loss. Surgical intervention for the arachnoid cyst was deferred, with spontaneous resolution at age two years without hearing recovery. CMA revealed a novel, de novo 5.1 Mb microdeletion of 22q13.31q13.33 not involving SHANK3, a gene typically deleted in PMS.

As diagnostic sensitivity improves, smaller chromosomal imbalances will be detectable related to milder or different phenotypes. They present two patients with novel deletions of chromosome 22q13 associated with multiple congenital anomalies and features distinct from PMS.

2016

A 14-year-old previously healthy girl presented to our outpatient clinic with a 6-weeks history of frontal headache. They typically would start in the occipital region and then radiate bifrontally. The neurological examination was unremarkable. Magnetic resonance imaging revealed an extra-axial bilateral lesion in bilateral cerebellopontine angle, larger on left side. The lesions were...
homogeneously hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging without evidence of contrast enhancement and without evidence of restriction on diffusion-weighted imaging. No surgical treatment was indicated.

Bilateral arachnoid cysts of the cerebellopontine angle are very infrequent and the main indication for surgery is the existence of clinical symptoms or neurological deficit coincident with the locations of the cysts 25).

2015

Petscavage et al. present the case of a 49-year-old woman who presented with acute, nonprogressive left sensorineural hearing loss and benign positional vertigo that was associated with an arachnoid cyst of the cerebellopontine angle. The presence of the lesion was documented by MRI examinations that were obtained 7 years apart. Arachnoid cysts at the cerebellopontine angle are usually found incidentally on MRI performed for unrelated reasons. However, if the arachnoid cyst displaces or compresses adjacent cranial nerves, symptoms may result. They review the salient imaging features of arachnoid cysts that allow their differentiation from other lesions of the cerebellopontine angle 26).

Gurkas et al. report a patient with a CPA arachnoid cyst. He presented with cranial nerve palsies and mirror movements found in upper extremities. They postulated that CPA arachnoid cyst compressing the brain stem and the pyramidal decussation may lead to mirror movements, and conclude that mirror movements can be associated with CPA arachnoid cyst 27).

A 71-year old woman presenting with a right hemifacial spasm and an ipsilateral arachnoid cyst. Preoperative magnetic resonance imaging findings suggested a neurovascular compression caused by displacement of the facial-acoustic complex and the anterior inferior cerebellar artery by the cyst. Cyst excision and microvascular decompression of the facial nerve achieved permanent relief. The existing cases of arachnoid cysts causing hemifacial spasm are reviewed and the importance of a secondary neurovascular conflict identification and decompression in these cases is highlighted 28).

Trigeminal Neuralgia in a Child With a Cerebellopontine Angle Arachnoid Cyst 29).

Sharma et al. report two cases of bilateral CPA AC with their pathophysiology and review of literature 30).

2014

Visagan et al. first report a CPA arachnoid cyst causing TGN in a paediatric case 31).

2012

A 62-year-old man complaining of vertigo and progressive hearing loss was diagnosed with an arachnoid cyst at the right cerebellopontine angle based on magnetic resonance imaging (MRI). In this case-report, we used computed tomography (CT) cisternography to determine whether the

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arachnoid cyst communicated with the cerebrospinal fluid (CSF) space. Differentiating between a noncommunicating and communicating arachnoid cyst is required for presurgical evaluation. CT cisternography is a less used but reliable radiological technique for determining the presence of communication, and could therefore be included in the diagnostic work-up of arachnoid cysts. The patient underwent surgery with fenestration of the arachnoid cyst; his vertigo improved and his hearing was preserved [32].

Superior oblique myokymia (SOM) is a rare disorder with an unclear pathogenesis. We describe a first reported case of chronic disabling SOM in association with a cerebellopontine angle arachnoid cyst, who had a gradual and eventually complete symptomatic resolution 8 months following cyst marsupialisation. Among other aetiologies, SOM may therefore be due to abnormal CSF flow dynamics resulting in structural compromise of the nerve [33].

2011

A 7-month-old infant presented to the hospital with a history of delayed milestones and an abnormal increase in head circumference. Magnetic resonance images and CT scans of the brain showed a large CSF cavity involving the entire brainstem and a right CPA arachnoid cyst causing obstruction of the fourth ventricle and dilation of the lateral and third ventricles. Cerebrospinal fluid diversion was performed by direct communication from the syringobulbia cavity to the left lateral ventricle and from the left lateral ventricle through another ventricular catheter; external ventricular drainage was performed temporarily for 5 days. Communication between the syrinx and arachnoid cyst was confirmed. Clinically, there was a reduction in head circumference, and serial MR imaging of the brain showed a decrease in the size of the syrinx cavity and the ventricle along with opening of the normal CSF pathways. The postoperative course was uneventful, and no further intervention was necessary. On follow-up of the child at 3 years, his developmental milestones were normal. Surgical intervention for this condition is mandatory. The appropriate type of surgery should be performed on the basis of the pathophysiology of the developing syringobulbia [34].

A 47-year-old woman complaining of sharp and lancinating pain in the right periauricular and submandibular areas visited our hospital. Swallowing, chewing, and lying on her right side triggered the pain. Her neurologic examination revealed no specific abnormalities. The results of routine hematologic and blood chemistry studies were all within normal limits. Carbamazepine and gabapentin were given, but her symptoms persisted. Her pain was temporarily relieved only by narcotic pain medication. MRI showed an arachnoid cyst located in the right cerebellomedullary cistern extending to the cerebellopontine cistern. Cyst removal was performed via a right retrosigmoid approach. Lateral suboccipital craniotomy was performed using the right park-bench position. After opening the dura and cerebellopontine angle, the arachnoid cyst was exposed. The arachnoid cyst was compressing the flattened lower cranial nerves at the right jugular fossa. Her symptoms resolved postoperatively. Two months after the operation, she was completely free from her previous symptoms [35].

2009

A rare case of cerebellopontine angle arachnoid cyst manifesting as hemifacial spasm (HFS) is reported. The patient is a 42-year-old woman with 10-month history of left HFS. A preoperative
magnetic resonance imaging scan showed a well-demarcated area, hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging, in the left cerebellopontine angle, without contrast enhancement, resembling an arachnoid cyst. METHODS: The cyst was excised with microneurosurgical technique and the facial, vestibular, and acoustic nerves were completely decompressed from the arachnoid wall. RESULTS: The postoperative course was uneventful, and the left HFS disappeared immediately. Histologically, the cyst wall was a typical arachnoidal membrane. Ten months after surgery, the patient is symptom free. CONCLUSION: It is well-known that in approximately 10% of cases, trigeminal neuralgia can be caused by a space-occupying mass. However, the fact that HFS can also be caused by organic lesions as well as neurovascular compression is less well-known. Although the occurrence of tumor compression causing HFS has been previously recognized, cerebellopontine angle cysts have very rarely been described. The observation of a patient with a cerebellopontine angle arachnoid cyst causing HFS prompted us to review the literature relative to HFS caused by an organic lesion rather than neurovascular compression. 

2007

a patient with a CPA arachnoid cyst who presented with hoarseness (unilateral vocal cord paralysis) and dysphagia secondary to isolated compression of the vagus nerve. This rare presentation of a CPA arachnoid cyst has not been reported previously. CLINICAL PRESENTATION: The patient described is a 50-year-old man who experienced a precipitous onset of hoarseness and dysphagia. An otolaryngological evaluation revealed right-sided vocal cord paralysis. Brain magnetic resonance images displayed a cystic mass at the right CPA and anterior displacement of the vagus nerve. INTERVENTION: The patient underwent retrosigmoidal craniectomy with cyst fenestration, which was well tolerated. Intraoperatively, Cranial Nerve X was found splayed over the cyst and was consequently decompressed. CONCLUSION: Postoperatively, the patient's dysphagia completely resolved. However, the results of a laryngeal electromyocardiogram revealed minimal evidence of recovery in the affected vocal fold, and the patient continued to suffer from dysphonia. Although CPA arachnoid cysts are rare, they should be considered when a patient presents with an isolated cranial nerve palsy. Treatment options include cyst fenestration and cranial nerve decompression.

2006

A 51 years old female is reported who was diagnosed by IRM of a 4.5 x 2 cm arachnoid cyst, situated on the left cerebello-pontine angle, with tinnitus, hearing loss and vertigo that mimicked a Meniere's attack. We think these benign tumors must be included in the differential diagnosis of Meniere's disease because they can be indistinguishable from it clinically.

1992

Higashi et al., reported the first case of hemifacial spasm with an ipsilateral cerebellopontine angle arachnoid cyst in a 25-year-old man. The patient underwent evacuation of the arachnoid cyst by a partial membranectomy without any beneficial effect, and finally got rid of the hemifacial spasm by reexploration and microvascular decompression of the facial nerve. The operative findings and results revealed that the cyst produced deviation of the ipsilateral posterior inferior cerebellar artery, which was secondarily in contact with the root exit zone of the facial nerve.

A 17-month-old girl who developed two cerebellopontine angle arachnoid cysts after posterior fossa surgery for a brain tumor. After surgical excision of the tumor the child developed a left cerebellopontine angle cyst. This was treated through a suboccipital craniectomy by evacuating the cyst and excising the cyst wall. Two months later the child developed a second right-sided...
cerebellopontine angle cyst. It was treated by inserting a cystoperitoneal shunt. This article presents the case with radiological evidence of the acquired nature of the cysts. It also includes a brief review of the clinical presentation, pathogenesis, radiological evaluation, and surgical treatment of arachnoid cysts with emphasis on those occurring in the posterior fossa.

1991

A case of an arachnoid cyst in the cerebellopontine angle manifesting as contralateral trigeminal neuralgia is presented. Decompression and excision of the lesion resulted in total relief of symptoms. The possible causes of contralateral trigeminal neuralgia are briefly reviewed, and the surgical treatment of this entity is discussed.

1987

A case of a cerebellopontine angle arachnoid cyst spontaneously disappeared is reported. A 1-year-and-11-month old boy was suffered from sudden onset of left facial palsy. CT scan demonstrated dilatation of left internal auditory canal and a cystic lesion in the left cerebellopontine angle. Neurological examination disclosed only left facial palsy and left hearing loss. There was no signs and symptoms of increased intracranial pressure. He was followed up by CT scan. Repeated CT scan showed non-enhanced cystic lesion, the attenuation value of which was similar to that of cerebrospinal fluid. The cyst expanded gradually, and the brain stem was severely compressed. Then operation was planned under the diagnosis of left cerebellopontine angle arachnoid cyst about 2 years after the onset. But CT scan performed before operation showed disappearance of the cyst. Without operation the patient was followed by CT scan. There is no recurrence of the cyst. Natural history of arachnoid cyst will be well understood with repeated CT scan.

References


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