

ARACHNOID CYSTS OF SYLVIAN FISSURE IN PEDIATRIC POPULATION – DIFFICULTIES IN QUALIFICATION FOR SURGICAL TREATMENT

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Abstract

Most arachnoid cysts (AC-arachnoid cysts) are detected accidentally and usually remain clinically silent.

When they rarely present with symptoms, these are usually: signs of acute intracranial hypertension or the full spectrum of chronic or intermittent symptoms (headache, seizures, psychomotor retardation etc.). Cause-effect in the case of the latter presentation is still a matter of debate and uncertainty.

While in cases of acute intracranial hypertension surgery is necessary and lifesaving due to the severe condition of the patient and can be confirmed by imaging studies, determining indications for surgery in the case of chronically elevated intracranial pressure is usually very difficult. Firstly, because of the wide variety of clinical symptoms, they do not represent a direct threat to the life of the patient. Secondly, the symptoms do not necessarily result from the cysts, they can be coincidental. Thirdly, brain CT and MR images are usually normal apart from the presence of an AC.

In this paper we would like to present our own experience in 3 cases of Sylvian fissure arachnoid cysts in children diagnosed and treated at the University Children's Hospital in Krakow.

Keywords: Arachnoid cysts, subdural hematoma, brain global ischemia, intracranial hypertension

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Introduction

An arachnoid cyst (AC-arachnoid cysts) refers to an abnormal collection of fluid contained between two layers of arachnoid membrane. The frequency of AC in children is estimated at 2.6% and this number increases proportionally to an increase in the availability and frequency of imaging studies performed due to various complaints [2,4]. Other sources state that arachnoid cysts represent 1% of all intracranial masses [13,26,27].

Depending on the level of communication between the lumen of the AC and the subarachnoid space we classify them as completely communicating, partially communicating and non-communicating [1,23].

The most common location of AC (approx. 60%) is the medial cranial fossa – a cyst of the Sylvian fissure. Less common are posterior fossa cysts, cysts of the convexity of the brain, suprasellar cysts and cysts of the interhemispheric fissure. Least common are cysts of the quadrigeminal

plate and cysts within the ventricular system [1,25,26].

An ACs of the Sylvian fissure are divided by Galassi into three types according to size.

Sylvian fissure type I arachnoid cysts, due to the broad communication with the basal cisterns and thus their small size and lack of clinical symptoms, do not require surgery.

Type III arachnoid cysts are large fluid spaces filling the medial cranial fossa, they invariably cause compression and displacement of the temporal and frontal lobes as well as of the longitudinal cerebral fissure. This results in symptoms of acute intracranial hypertension and so require surgical treatment.

The most problematic group are those patients with Galassi type II arachnoid cysts; in these cases, even if complicated by haemorrhage, the patient is usually clinically stable and conservative treatment is viable. This is consistent with the ability of some arachnoid cysts to be self-limiting,

as in the second case described in our paper.

On the other hand, some authors maintain that once damaged, blood vessels of the cyst wall are more prone to recurrent bleeding as a result of even minor trauma. Recurring haemorrhages into the arachnoid cyst and subdural space cause decreased resorption of cerebrospinal fluid which results in enlargement of the cyst and the formation of a chronic subdural haematoma.

The vast majority of ACs remain clinically silent.

Other cases of symptomatic ACs result either from acute intracranial hypertension or from chronically elevated intracranial pressure, which can manifest itself in a very wide spectrum of symptoms such as headaches, seizures, vomiting, visual disturbances, neuropsychological symptoms, cranial nerve palsies, paresis and endocrine disorders [4,8,9,10,11,12,15,16,17,18,21,22,24]. The last occur in suprasellarly located ACs. Often it is difficult to establish a link between the presence of arachnoid cysts and the symptoms reported by the patient, which may or may not necessarily result from chronically elevated intracranial pressure. According to some authors an arachnoid cyst may only aggravate existing problems. This implies difficulty in making a decision as to their treatment, especially whether to undertake neurosurgery, which carries the risk of serious complications.

A separate issue are arachnoid cysts in which there have been complications such as bleeding into the cyst lumen and / or subdural space, usually after minor trauma. The mechanism of this phenomenon is not fully understood. One theory is that the AC is less able to withstand traumatic forces than normal brain tissue. The pressure that is created in light of the cysts, even in minor trauma, is transmitted to the outer wall causing tearing of the small blood vessels. In such cases, a sudden increase in the size of the cyst or an increase in pressure within it and the appearance of subdural hematoma lead to acute intracranial hypertension necessitating life-saving neurosurgery [13].

Avoidance of this complication is presented as an argument for prophylactic surgical treatment. Those opposed to such early surgical intervention argue that ACs may undergo spontaneous „self-cure“ when the cyst wall ruptures (without meaningful vascular damage) evacuating its contents into the subdural space with subsequent hygroma formation. These will then sometimes

be resorbed. Both sides of the discussion, therefore, use the same mechanism, i.e.: cyst wall breakage, as the basis for their argument [7,19,20].

We present three cases of ACs – the first two feature acute intracranial hypertension with subdural haemorrhage and the third symptoms suggestive of chronic elevated intracranial pressure. The first case caused diagnostic difficulties during the early phase due to an atypical presentation in CT and MRI. The second case is a typical presentation of a ruptured arachnoid cyst, with features of acute intracranial hypertension, which on the basis of radiological imaging and clinical symptoms was qualified for surgery; however, due to the lack of parental consent for the procedure conservative treatment was implemented. The third of the presented cases is a patient whose symptoms could be due to the presence of chronic intracranial hypertension caused by the presence of an arachnoid cyst. This cyst did not present with radiological evidence of intracranial hypertension. The patient remains in pharmacological treatment.

Case presentation

Case 1

This 2-year-old girl was admitted to the University Childrens Hospital with very severe headache on admission having had a short-term loss of consciousness. History presented a few days of headache and with morning vomiting.

At the age of 7 months she was seen by a neurologist for suspected epilepsy, which ultimately was not confirmed in further clinical examination and observation.

Clinical examination at admission showed positive bilateral Babinski reflexes. No other abnormalities were found.

An urgent CT scan (without intravenous contrast) of the head was performed, which revealed an isodense area 10mm wide adjacent to the brain in the right fronto-parietal area (Fig.1). Fresh blood was also found in the Sylvian fissure, along the cerebellar tentorium and in the anterior part of the falx cerebri as well as edema of the right hemisphere, with narrowing of the cerebral sulci and fissures (including the Sylvian fissure) and blurring of the cortico-subcortical border (Fig. 2).

The zone described above could suggest the presence of subacute subdural hematoma, but the source of bleeding, based on CT scans, remained unclear.

A differential diagnosis of trauma was taken into account but the parents negated any traumatic injury in the previous weeks. A CT scan also did not confirm the presence of other post-traumatic changes such as fractures of the skull, the presence of hematoma and / or edema in the soft tissues of the epicranial aponeurosis. Hematological disorders were also excluded.

Bleeding as a change secondary to a pre-existing defects or vascular lesion such as vascular malformation or aneurysm were considered – however the study was performed without intravenous contrast agent due to the serious condition of the child and the possibility of toxic brain damage due a damaged blood-brain barrier. Also, the location of the hematoma was not typical of a ruptured malformation or aneurysm. Another potential cause taken into account was complications such as bleeding from a ruptured AC but no AC was found in the performed CT scan (the girl had had no previous imaging studies of the head).

Other, much less probable hypotheses were : the presence of purulent exudate (at time of performing the CT no inflammation of the CSN had been confirmed or excluded) or meningeal tumor with secondary bleeding.

MRI performed the next day confirmed the presence of subdural hematoma on the right, but

the image was untypical in the context of the presented history (predominantly isointensive in T1 and T2 with small areas hyperintensity in T2).

In addition, edema of the right hemisphere with subfalcine herniation was found (Fig 3,4).

In conclusion, the MRI image revealed the presence of a subdural hygroma-hematoma, which most likely resulted from a ruptured arachnoid cyst of the Sylvian fissure – which in the performed MRI was already moderately expanded.

The progression of symptoms suggestive of subfalcine herniation, deterioration of the patient's GCS score from 15 points at admission to 13 points and because the CT and MRI images revealed features of acute cranial hypertension, it was decided that surgery was necessary.

A right fronto-parieto-temporal craniotomy was performed, a hemolized subdural hematoma was evacuated and its' outer wall removed. Intraoperatively within the enlarged Sylvian fissure blood and fragments of arachnoid containing venous vessels were found, confirming that the cause of bleeding was a ruptured AC.

During hospitalization, after the evacuation of the hematoma, follow-up CT scan of the head was performed which revealed a subdural hematoma, narrower than before the operation. Compression of the right hemisphere of the brain and the right



Fig. 1. Nonenhanced CT axial scan



Fig. 2. Nonenhanced CT axial scan

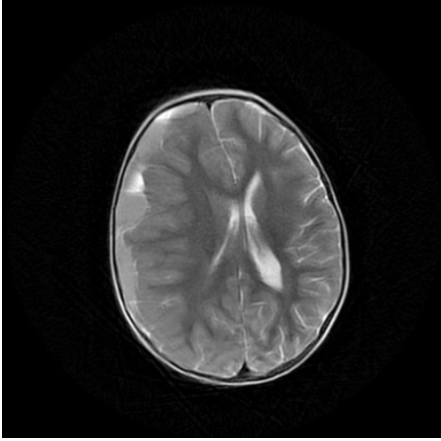


Fig. 3. T2WI axial image

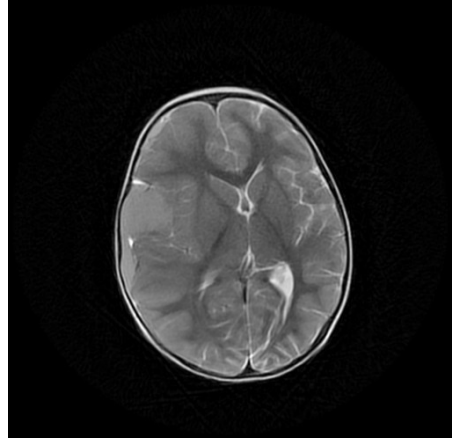


Fig. 4. T2WI axial image

lateral ventricle were also markedly reduced. Right hemispheric edema was also smaller with clearly visible differentiation of the cortico-subcortical border. In addition the presence of arachnoid cysts (Galassi type II) were found.

In the postoperative period there was a subaponeurotic leak of cerebrospinal fluid – a lumbar-peritoneal drain was implanted resulting in an improvement of the the local and overall neurological condition. The patient was discharged home with no meningeal symptoms and without signs of focal damage to the CNS. There was no clinical or radiological evidence of intracranial hypertension.

Case 2

This 7-year-old girl was admitted because of headaches increasing in intensity over several days, and abnormal vision in the right eye. Two days before admission she started vomiting. The symptoms had begun following a head injury as a result of a fall on ice three weeks previously.

An urgent CT scan of the head was performed, which revealed a cyst on the right side of the Sylvian fissure (type II according to Galassi) with expansion of the fluid space temporo-parietally adjacent to the brain congruous for a hygroma containing a trace of blood. In addition, the sign of compression by a mass was found in the form of displacement of the longitudinal cerebral fissure to the left and right lateral ventricle

compression as well as slight local edema with smoothing of the cerebral sulci of the right hemisphere (Fig. 5,6). CT imaging suggested rupture of arachnoid cysts most likely due to trauma, followed by acute intracranial hypertension.

The ophthalmic examination found a blurred edematous optic nerve disc in the right eye. Antiedema medication was started and the patient qualified for surgery. The child's mother did not consent to surgery, the girl was therefore treated medically. After a few days of hospitalization the patient was discharged home at the request of the mother. On the day of discharge clinical signs of intracranial hypertension were absent and there were no symptoms of focal neurological damage. A few days after the CT scan a follow-up MRI of the head was performed. The image of the brain was comparable. Both longitudinal cerebral fissure displacement and right lateral ventricle compression were still present (Fig. 7,8).

The child remains under neurosurgical outpatient care – during a few months of observation the child's clinical condition remains good, with no clinical signs of intracranial hypertension. The control MRI scan carried out after 8 months showed complete regression of the hygroma on the right side, with the AC still present – a little bit bigger than previously. No signs of cerebral oedema or longitudinal fissure displacement were found (Fig 9,10).

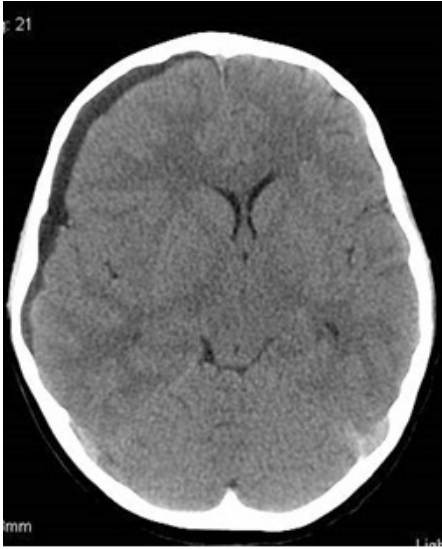


Fig. 5. Nonenhanced CT axial scan

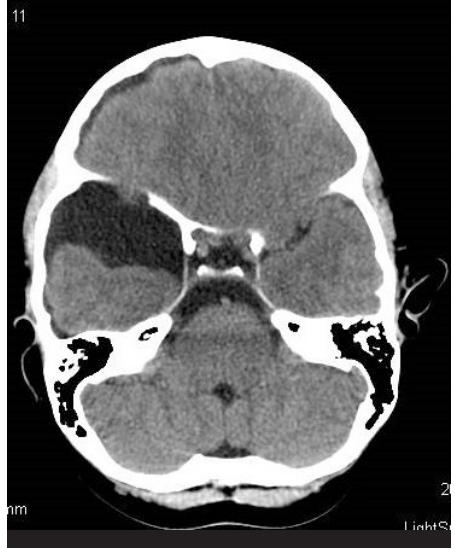


Fig. 6. Nonenhanced CT axial scan

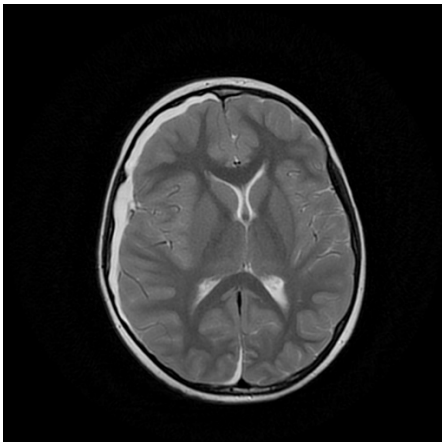


Fig. 7. T2WI axial image

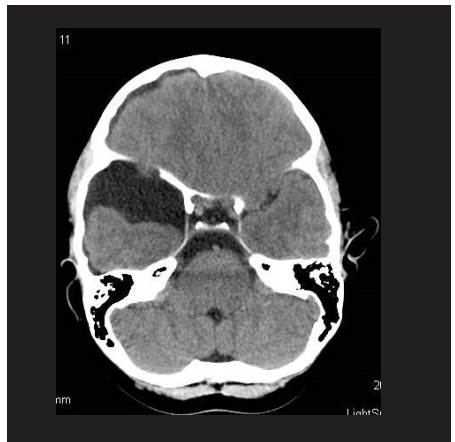


Fig. 8. T2WI axial image

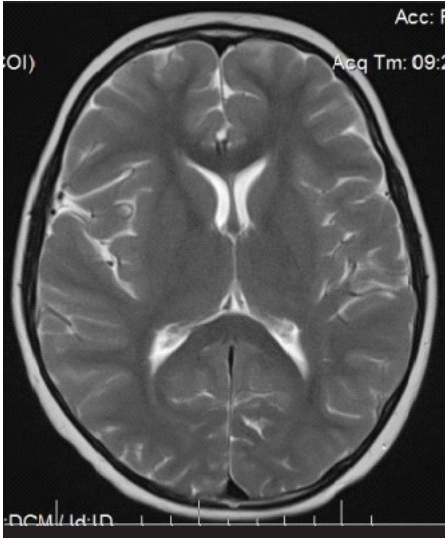


Fig. 9. T2WI axial image

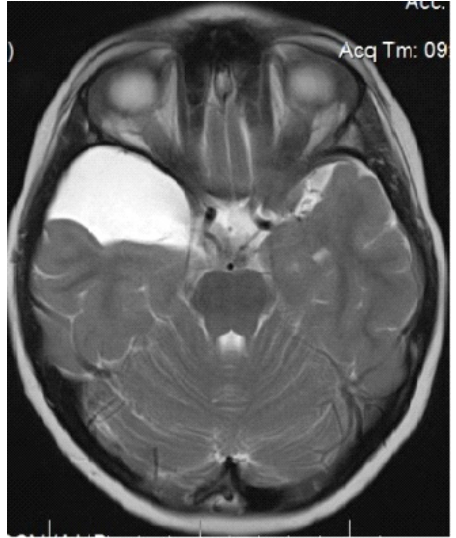


Fig. 10. T2WI axial image

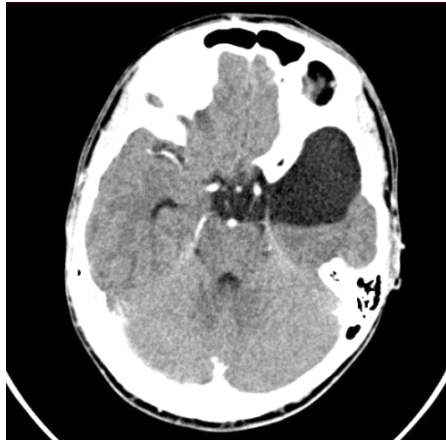


Fig. 11. Contrast-enhanced CT axial scan

Case 3

This 14 year-old boy was admitted to the University Childrens' Hospital after a week of mental disorders in the form of arousal, dissociation, difficulty in falling asleep, periodically feelings of "being outside his body." A few days later those symptoms werejoined by illusions in the form of micropsia and makropsia: some items seemed to the patient to be bigger than in reality, others smaller. The boy remained critical of these visual experiences which ruled out a psychotic disorder, and rather pointed to an organic cause of his reported symptoms. For this reason, a CT scan of the head was performed, which revealed an arachnoid cyst of the left side of the Sylvian fissure – Galassi type II (Fig.11). There were no radiological signs of intracranial hypertension. Neuropsychological assessment revealed normal intellectual development and an average level of intelligence.

The boy was suspected of temporal lobe epilepsy but EEG didn't confirm the diagnosis. Pharmacological treatment (Carbamazepinum) was implemented, which significantly improved the clinical condition of the patient – reported symptoms disappeared almost completely. After a few months, in spite of treatment, the aforementioned symptoms recurred, although they were decidedly less severe. The boy remains under observation.

Discussion

It is widely assumed that the majority of arachnoid cysts remain asymptomatic, while few cases with symptomatic arachnoid cysts, which according to current knowledge are eligible for surgery, are patients with clinical and radiological features of acute intracranial hypertension.

The most problematic group of patients are those with symptoms which , although not life-threatening, evidently cause impaired quality of life; these patients usually have Galassi type II ACs.

In practice, the neurosurgeon must answer the following questions:

are the patients' symptoms caused by the AC or are the symptoms coincidental?

Is the presence of the AC causing exacerbation of the symptoms of a different disorder ?

In search of the answers to these questions medicine is turning to methods of assessment of brain function (SPECT, perfusion MRI/CT and functional MRI) rather than just brain anatomy,

which, as mentioned earlier, is usually normal other than the presence of the AC itself [3,7,14].

In our opinion this is the correct path to take, although present techniques, while showing certain pathological changes, still cannot provide us with definite answers as to the desired form of treatment to implement. One must remember that some of these imaging methods require application of significant doses of radiation (SPECT, PET, CT) so one must remember that „the biggest contrindication to the use of X-rays is a lack of indications” – especially in children.

Sgoruos and Chapman published a study of functional brain imaging of three children aged 3, 6 and 16 years old [16]. Two of them complained of periodic severe headaches suggestive of rupture of arachnoid cysts, and one was suspected of and diagnosed in the direction of autism.

These childrens development was normal and on neurological examination they also presented no deviations from the norm. Ophthalmic examination showed was no swelling of the optic nerve disc.

CT and MRI , despite the presence of arachnoid cysts in the Sylvian fissure did not reveal any changes characteristic of hypertension or intracranial bleeding.

In children functional SPECT CT scans were performed which in all three cases showed features of generalized reduced cerebral blood flow, not only in the hemisphere with arachnoid cysts (which can be explained by local compression of blood vessels by the cysts), but also in the opposite hemisphere , comparable to that in patients with intracranial hypertension caused by eg. hydrocephalus.

The patients underwent neurosurgical partial removal of the cyst wall. In all cases a short time postoperatively the patients' subjective symptoms of headache resolved. Control SPECT CT after surgery showed normal perfusion as opposed to that before surgery. The children's neurological condition after surgery and long-term observation did not raise any concerns. The thesis put forward by the authors suggests that even seemingly asymptomatic cysts cause signs which can only be radiologically found by advanced imaging techniques [16].

The validity of this conclusion would seem to confirm research carried out by Di Rocco et al involving prolonged (48 to 72 hours) measurement of the intracranial pressure in the region of cysts of the Sylvian fissure. They showed constantly

raised pressure in the case of type III arachnoid cysts and normal pressure in children with type I cysts.

The most difficult to interpret were the results of intracranial pressure testing in children with Galassi type II ACs; In half of them pressures were normal throughout most of the testing period while the other half demonstrated elevated pressure levels [6].

Although the problem of arachnoid cysts has been frequently addressed in medical literature, the criteria for surgical intervention in a number of these cases remain uncertain.

Case 3 described by us illustrates this problem – the issues related to the presence of chronically or periodically increased intracranial pressure due to the presence of AC.

It may be the cause of reversible reduced perfusion and metabolism of the brain and it this which is most likely responsible for the symptoms reported by patients. However, a clear demonstration of cause and effect is often very difficult or even impossible.

The question about which method of treatment to use therefore remains open, because the indications for surgery are inconclusive.

Over the last few years in our hospital our approach has changed, with an evident shift away from prophylactic surgical treatment and towards conservative treatment.

We must also remember that surgery in patients with chronically or periodically increased intracranial pressure aims „only” to improve their quality of life and is not a life-saving procedure as in the case of acute intracranial hypertension. Last but not least, when considering the pros and cons of possible surgical intervention one cannot forget about the possible complications of neurosurgery.

Resumo

La plejmulto de araknoidaj cistoj (AC-oj) estas trovitaj hazarde kaj kutime restas klinike mutaj. Kiam ili malofte kaŭzas simptomojn, tiuj ĉi kutime estas: signoj de akuta intrakrania hipertensio aŭ la tuta aro de kronikaj kaj kelkfojaj simptomoj (kapdoloro, iktoj, psikomotoraj malrapidiĝo k.t.p.). La rilato inter kaŭzo kaj efiko okaze de la lasta sinprezento estas daŭra temo de debatoj kaj necerteco. Malgraŭ la fakto, ke je kazoj de akuta intrakrania hipertensio operacio estas necesa kaj vivsavanta kaŭze de la malfavora situacio de la paciento, kio povas esti konfirmita per bilddonantaj ekzamenmetodoj, kutime ege malfacilas certigi indikacion por operacio

kaze de kronike altigita intrakrania premo. Unue, kaŭze de la granda diverseco de klinikaj simptomoj, tiuj lastaj ne rekte minacas la vivon de la paciento. Due la simptomoj ne nepre rezultas de la cistoj, sed povas esti koincidaj. Trie, kutime komputila tomografio kaj magneta resonanca bildigo estas normalaj pro la ekzisto de AC. En tiu ĉi artikolo ni prezentas nian propran sperton kun 4 kazoj de AC-oj de la Silvia fisuro je infanoj, kiuj estis diagnozitaj kaj kuracitaj en la Universitata Infanohospitalo de Krakovo.

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