Clinical case

Adult symptomatic and growing arachnoid cyst successfully treated by ventriculocystostomy: A new insight on adult arachnoid cyst history

Kyste arachnoïdien symptomatique et évolutif de l'adulte traité par ventriculocystostomie : un nouveau regard sur l'évolutivité des kystes arachnoïdiens de l'adulte

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ABSTRACT

Background. – Adult arachnoid cysts are known to be stable and asymptomatic but their history remains undefined.
Case description. – The authors report the case of an 81-year-old woman with progressive hemiplegia and aphasia. CT scan revealed a voluminous left frontotemporal arachnoid cyst with a major mass effect on the midline and contralateral blocked hydrocephalus. Endoscopic ventriculocystostomy was performed with a spectacular neurological improvement.
Discussion and conclusions. – Symptomatic adult arachnoid cysts are extremely rare. To our knowledge, no similar clinical case of a growing arachnoid cyst in elderly patients has yet been reported in the literature. The mechanisms of cyst enlargement and decompensation still remain undefined and debated. The possibility of adult arachnoid cyst growth has to be considered in clinical practice. Endoscopic ventriculocystostomy is as effective as in paediatric cases.

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RéSUMÉ

Cas clinique. – Les auteurs rapportent le cas d’une patiente de 81 ans présentant une hémiplegie et une aphalie d’aggravation progressive. Le scanner cérébral réalisé mettait en évidence un volumineux kyste arachnoïdien frontotemporal gauche avec un important effet de masse sur les structures médianes ainsi qu’une hydrocéphalie bloquée contro-latérale. Une ventriculocystostomie par voie endoscopique a été réalisée permettant une amélioration neurologique spectaculaire.
Discussion et conclusions. – Les kystes arachnoïdiens symptomatiques de l’adulte sont extrêmement rares. À notre connaissance, on ne retrouve pas dans la littérature de cas de kyste arachnoïdien évolué chez une patiente aussi âgée. Les mécanismes expliquant l’augmentation de volume des kystes arachnoïdiens demeurent incertains et débattus. L’éventualité d’une augmentation de volume d’un kyste arachnoïdien de l’adulte doit être prise en compte en pratique clinique. Dans cette indication, la ventriculocystostomie par voie endoscopique est aussi efficace chez l’adulte qu’en pédiatrie.

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1. Background

Arachnoid cysts are frequent and well studied in paediatric diseases, but rarely occur in adults. Adult forms are known to be stable and asymptomatic, and therefore, neurological follow-up is not
required [1–3]. Nevertheless, the history of adult arachnoid cysts remains undefined.

The authors report the case of an 81-year-old woman with symptomatic and growing frontotemporal arachnoid cyst that was revealed by a quickly progressive aphasia and hemiplegia.

2. Clinical presentation

The authors report the case of an 81-year-old woman referred to the neurological emergency unit after she experienced a 48 h-history of aphasia and hemiplegia. A two-year history of cognitive impairment was reported that led to the prescription of donepezil the previous year due to a diagnosis of supposed senile dementia. A CT scan performed one year earlier revealed a left paraxial supratentorial frontotemporal cyst, which was not suspected at that time to be involved in the patient’s symptoms (Fig. 1A).

Clinical examination at admission revealed aphasia with complete right hemiplegia. The Glasgow Coma Scale was 8. Cerebral CT scan performed at admission showed a voluminous left frontotemporal cyst with an important mass effect on the midline and lateral ventricles, associated with a contralateral blocked hydrocephalus (Fig. 1B and C). Cyst and cerebrospinal fluid densities were equivalent on pre-contrast CT scan. Post-contrast CT scan did not show any contrast enhancement or any argument for a tumour mass. The main hypothesis was an arachnoid cyst. Moreover, a comparison with the previous year’s CT scan showed an obvious increase in the cyst volume with contralateral blocked hydrocephalus and peri-ventricular oedema.

Subsequently, we decided to perform an endoscopic marsupialisation of the cyst located in the left lateral ventricle. Ventriculocystostomy (VCS) was performed using a catheter balloon. Macroscopic cyst appearance was typical of an arachnoid cyst. Endoscopic cyst exploration did not show any tumoral lesion. Cyst content and CSF biology were the same and normal. Postoperative cerebral magnetic resonance imaging (MRI) confirmed ventriculocystostomy patency (Fig. 1D).

Postoperative neurological improvement was spectacular with complete aphasia recovery and progressive right motor improvement. Five days after surgery, the patient was discharged and able to walk with help. Three months later, the patient was at home and had recovered complete autonomy. Follow-up CT scans showed a decrease in the cyst volume and mass effect on medial structures, with no residual hydrocephalus or peri-ventricular oedema (Fig. 1E and F). Two years later, neurological examination was still normal.

3. Discussion and conclusion

Symptomatic adult arachnoid cysts are extremely rare [1,3,4]. To our knowledge, no cases of growing symptomatic arachnoid cyst in patients, who are elderly, have ever been reported in the literature, highlighting the originality and interest of this particular case. Al-Holou et al., studying the prevalence and natural history of the paediatric arachnoid cyst, did not observe any cyst growth in children after four years of age and demonstrated a younger age to be significantly correlated to cyst enlargement [5]. Several cases of expanding arachnoid cysts have been reported in literature, but this systematically concerned the paediatric

![Fig. 1. Cerebral CT scan performed one year previously (A) showed less important mass effect on medial structures and no hydrocephalus compared to preoperative CT scan (B). Axial (B) and coronal (C) preoperative CT scan showed voluminous left para-axial frontotemporal arachnoid cyst with severe mass effect on medial structures and contralateral blocked hydrocephalus. Notice the absence of squamous temporal bone thickening. Early postoperative MRI (D) showed arachnoid cyst after ventriculocystostomy. Respective 3 months (E) and 1 year (F) postoperative CT scans showed a decrease in cyst volume and mass effect on medial structures with no residual contralateral hydrocephalus.](image-url)
disease. In their literature review, Rao et al. reported only 5 cases of documented arachnoid cyst growth in children aged 4 months to 8 years [2]. In the adult, Oertel et al. reported 11 cases of paraxial supratentorial symptomatic arachnoid cysts successfully treated by VCS [6,7]. The patients’ ages ranged from 14 to 71 years old. Talamonti et al. reported a series of 10 cases of cortical arachnoid cyst, including children and adults [8]. However, the cyst reported in these series was not so voluminous with a severe midline shift.

The diagnosis of an arachnoid cyst could be discussed. The absence of squamous temporal bone thickening is atypical in congenital arachnoid cyst and the precise physiopathology could be questioned. Unfortunately, no previous cerebral CT scan has been performed during the patient’s lifetime. The most frequent adult intracerebral cysts aetiologies are tumoral: metastases, gliomas and rare cases of glio-epithelial cysts. One case of adult intracerebral cyst has been described [9]. However, in this case, the CT scan and MRI (Fig. 1A–D) were typical of an arachnoid cyst, except for the absence of squamous temporal bone thickening [10]. Macroscopic aspect and cerebrospinal fluid analyses were equally characteristic of an arachnoid cyst. There was no argument for a tumoral or other origin and therefore, the diagnosis of an arachnoid cyst was retained. Nevertheless, the diagnostic hypothesis of Virchow-Robin space (VRS) dilation had to be considered. Giant VRS dilation is generally described to be localized in the mesencephalic or diencephalic area. However, VRS dilatations are equally observed in the convex cortex [11–13]. In these cases then, giant convexity VRS dilation origin could be suggested.

In this reported case, a comparison CT scan performed one year before and at admission interestingly showed evidence of cyst enlargement with increasing mass effect on adjacent structures. In this instance, the case would involve questioning arachnoid cyst history and physiopathology.

The mechanisms of cyst enlargement and decompenation remain undefined and debated [4,14,15]. Cyst growth is rare in paediatric cases of the disease and exceptional in adult. Different hypotheses, such as partial cerebral lobe agenesis, primary malformation of the arachnoid layer, mechanism of osmotic gradient between the intra- and extracystic medium, cyst membrane secretion have been previously suggested in paediatric arachnoid cyst with no evident certainty [1,16,17]. A hypothesis of sequestration by a ball-valve mechanism appears unlikely in this case due to the distance between the cyst and large vessels [18]. Physiopathology remains complex and a combination of these different hypotheses could be suggested. Post-traumatic cyst enlargement and intracystic bleeding have been suggested but no definite evidence has been proposed based on the clinical medical history, CT scan density and endoscopic appearance. Therefore, our case clearly documents adult arachnoid cyst growth and also provides new evidence as regards arachnoid cyst evolution.

Endoscopic VCS represents the reference treatment in child arachnoid cysts [14,15,19–21]. Oertel et al. and Talamonti et al. equally reported VCS efficacy in adult arachnoid cysts [6–8]. Cases reported in the literature, as well as series, are particularly rare and even non-existent in the elderly patient population. In this case, VCS appeared as the most logical surgical strategy. VCS efficacy was evident during the first two postoperative years despite the advanced patient age and the severity of the preoperative neurological status. In this case, the surgical procedure was uneventful. However, in some cases, localization of the VCS perforation point could be delicate, justifying neuronavigational use [6,7]. The risk of significant brain shift caused by opening cyst and content drainage also has to be considered. In cases of VCS failure, a cysto-peritoneal shunt is suggested.

In conclusion, the authors report a rare and original case of adult symptomatic and growing frontotemporal arachnoid cyst successfully treated by VCS, as well as providing a new insight into arachnoid cyst history. Also, the possibility of adult arachnoid cyst growth has to be considered in clinical practice. VCS appears as effective in the paediatric disease as well as in the elderly.

**Disclosure of interest**

The authors declare that they have no conflicts of interest concerning this article.

**References**


