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Supratentorial arachnoid cyst mimicking a Ménière's disease attack

Giuseppe Buongiorno, M.D., Giovanni Ricca, M.D.

Abstract
Arachnoid cysts (AC) often present with symptoms depending on their anatomical location within the skull; while supratentorial cysts grow causing relatively few symptoms, infratentorial ones may impair liquor circulation at the level of the fourth ventricle, giving rise to intracranial hypertension, or may stretch the complex nerve network in the cerebello-pontine angle. We report the singular clinical history of a 54-year-old male, who suddenly began to feel dizzy while sitting at his workplace, complaining of left tinnitus and aural fullness, in a classic clinical picture of Ménière's disease. The anomalous findings at otoneurological examination (markedly the left deviation at the Fukuda test) made a magnetic resonance image (MRI) scan mandatory and a huge AC was found in the left fronto-parietal lobe. The mass provoked an evident clockwise rotation of the brainstem that we suppose to be the cause of acoustico-facial bundle stretching explaining the vestibular symptomatology.

Key words: Arachnoid; Cysts; Ménière's Disease; Diagnosis, Differential

Introduction

Arachnoid cysts (AC), firstly described by Bright in 1831,[1] are fluid-filled spaces generated by congenital anomalous splitting of the arachnoid membrane; they may communicate with the subarachnoid space or may be partially or completely separated from it.

Arachnoid cysts may form in any position within the skull, more commonly in the left hemisphere, in the middle and anterior cranial fossae; in this region, arachnoid cysts may reach a large size, producing a marked displacement of the temporal and frontal lobes.

In a lower percentage of cases (from 20 to 30 per cent),[2] arachnoid cysts are located in the posterior cranial fossa, where they may occupy a retrocerebellar area, or grow in the cerebello-pontine angle. Signs and symptoms related to the presence of arachnoid cysts are somewhat different depending on their location: while supratentorial cysts may produce mild untreatable headache, difficulties in concentration, more rarely temporal lobe epilepsy and, in paediatric patients, bulging of parietal bone, posterior fossa arachnoid cysts tend to have a protean clinical manifestation, with hypoacusis, tinnitus, dysmetria, ataxia, sometimes vomiting and papilloedema from endocranial hypertension, from fourth ventricle obstruction.

In this article we report the singular history of a patient affected by a large supratentorial cyst that first manifested with signs and symptoms usually caused by posterior fossa expansile masses; furthermore, we try to explain the reasons for this symptomatology, by interpreting the MRI scans.

Case report

A 54-year-old executive male, came to Accident and Emergency complaining of acute peripheral vertigo, together with moderate nausea, tinnitus and fullness in his left ear, mimicking a Ménière's attack. The patient had begun to feel dizzy at rest, without any form of aura, while sitting at his workplace. In the months preceding this acute episode of vertigo, the patient had noticed a transient aural fullness, mostly localized in his left ear.

At our first observation, spontaneous horizontal second degree right beating nystagmus could be observed. The patient was given i.v. anti-emetic therapy and intravenous diazepam in slow infusion; this therapy was changed on the second day, with administration of i.v. 18 per cent glycerol, 500 ml a day.

On otoneurological examination, performed four days after admission, the patient showed no residual spontaneous nystagmus; on tandem Romberg test an evident left sway could be appreciated and there was a marked (>90°) left rotation on the Fukuda march test. On Dix-Hallpike manoeuvre, atypical positional vertigo and nystagmus were revealed: with the head bent to the left, pure horizontal apogeotropic nystagmus was elicited, resembling lateral canal positional vertigo. The pure tone audiometry showed a mild bilateral sensorineural hearing loss; on bi-thermal caloric tests, there was complete absence of response on stimulation of the left ear.

Because of these otoneurological findings; the patient was referred for a gadolinium-enhanced MRI scan. The examination showed a large ($10 \times 8 \times 7.5$) space-occupying mass, localized in the left fronto-temporal region. The mass, cystic and well rounded, was hypointense on T1-weighted scans and hyperintense on T2 scans, with no contrast enhancement and not communicating with the CSF spaces (Figure 1). The left parietal and temporal lobes were clearly compressed backwards (Figure 2), with moderate deformation of left lateral ventricle, but without clear CSF circulation impairment; the ventricular system

From the II Otolaryngologic Clinic, University of Turin, Turin, Italy.
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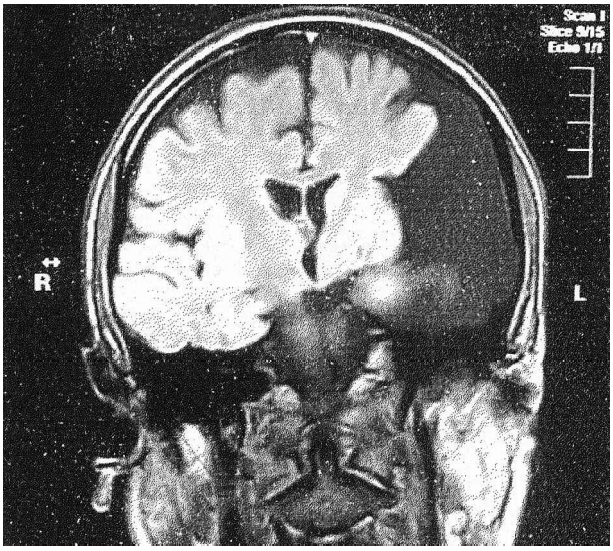

FIG. 1

Coronal T1 MRI scan showing a large arachnoid cyst replacing about half of the temporal lobe and deviating the contralateral cerebral hemisphere off the midline, with no enlargement of the cerebral ventricles.

showed diameters within normal ranges and there was no sign of parenchymal damage. Interestingly, some modification of normal anatomy was apparent in the posterior fossa: shift of the brainstem to the right was evident on coronal sections, together with clockwise rotation along its vertical axis, demonstrable on axial MRI scans (Figure 3). The patient was then referred to neurosurgeons for a further evaluation; to date no operation has been performed.

Discussion

Arachnoid cysts represent about one per cent of all intracranial lesions,[3] affecting more commonly the supratentorial area, in particular the Sylvian fissure; they are considered developmental in nature[4] and are diagnosed early in the course of life in 60 to 70 per cent of afflicted patients. The male/female ratio ranges from 2/1 to 3/1 in the different reports[5,6] and average duration of symptoms before the definitive diagnosis may vary from six months to five years.[6]

Adult arachnoid cysts represent about 20 to 30 per cent of the total cases and present from the third to the fifth decade of life, very rarely beyond 60. The sylvian fissure is by far the most common location for arachnoid cysts, especially on the left side; infratentorial cysts occur as well, usually causing symptoms related to intracranial hypertension, due to impairment of CSF flow through the fourth ventricle. Small cysts can be found accidentally on MRI or CT scan performed for other reasons; in adults these cysts may remain clinically silent for years, even for the whole lifespan, requiring only periodical MRI scanning. In other cases arachnoid cysts reach large diameters, especially when supratentorial; in some cases, an operation becomes mandatory even in aged patients, because of progression to dementia, hemiparesis and ataxia.[7]

In the majority of patients affected with AC the symptoms depend on the location and may have a long, vague and non-specific course, often lasting years. In some subjects, the presence of the arachnoid cyst may predispose the formation of subdural haemorrhage after mild cranial trauma[8] and the patient begins to complain of

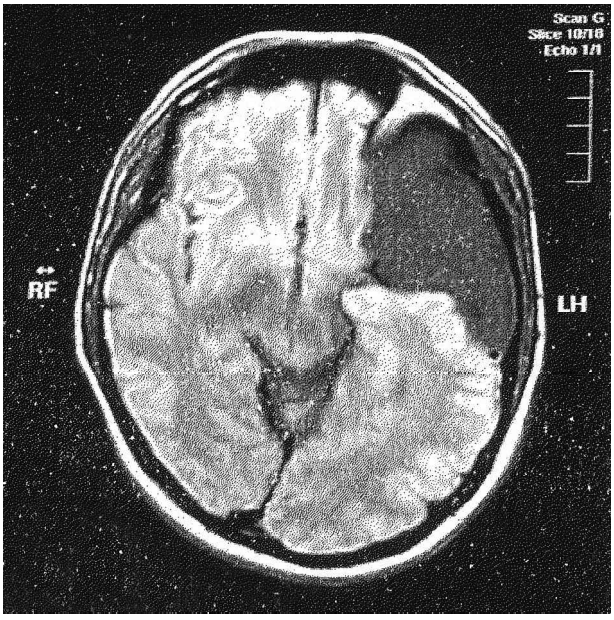

FIG. 2

Axial 'FLAIR' MRI scan demonstrating the anterior-posterior extension of the cyst and the evident posterior displacement of the whole left hemisphere; the region of 'torcular herophili' appears completely shifted toward the right side.

rapidly worsening symptoms. Complicated arachnoid cysts may present with progressive headache, nausea, vomiting, and double vision.

Intratentorial cysts are rarer (from 20 to 30 per cent of total), although the cerebellopontine angle remains the second most common location after the sylvian fissure.[9] Retrocerebellar, supracerebellar, clivar and vermian locations are also possible but are uncommon.

The most common symptoms related to posterior fossa arachnoid cysts are nausea, vomiting, headache and papilloedema, together with the cerebello-pontine angle syndrome.[10] Interestingly, vestibular symptoms (vertigo

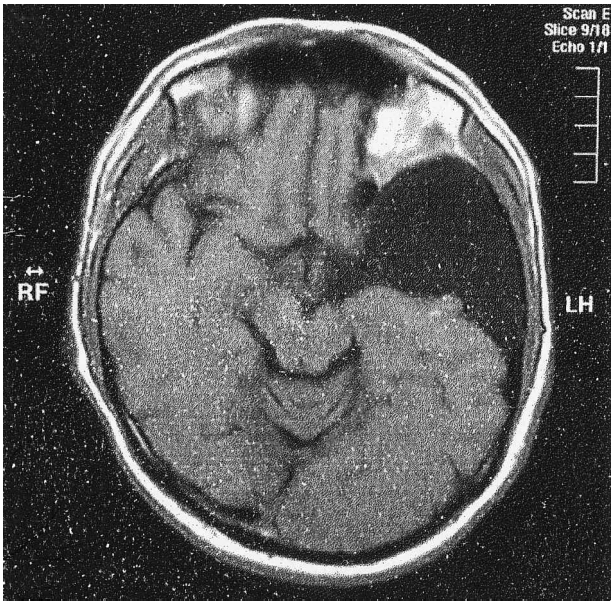

FIG. 3

Axial MRI scan at the level of middle crania fossa floor: notice that the left temporal lobe is substituted by the cyst and the ventral surface of the pons is splayed posteriorly.

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and dizziness) predominate over cochlear symptoms (tinnitus and hearing loss). Nystagmus is by far the most common clinical sign.

- **This is a case report of a patient who presented with symptoms similar to Ménière's disease**
- **Investigation showed the patient to have an arachnoid cyst**
- **At the time of reporting of the case surgery had not been undertaken**
- **It is postulated by the authors that the presentation is the result of compression of the VIIIth cranial nerve bundle**

It has been accepted for a long time that clinical presentation of arachnoid cysts depends on their location, except for symptoms due to intracranial hypertension; nevertheless, several papers (Haberkamp *et al.*,[11] McCullogh *et al.*,[12]) have described atypical presentation of arachnoid cysts, especially in case of posterior fossa location.

An atypical onset has been described by Jallo *et al.*[13] (1997), including ataxia, dysdiadokinesia, giddiness (cerebellar) and reduced corneal sensitivity.

Babu and Murali[14] described an arachnoid cyst presenting with contralateral trigeminal neuralgia, while Higashi and colleagues[15] reported a 25-year-old male whose initial complaint was ipsilateral hemifacial spasm. In both these cases, the arachnoid cyst was located in the posterior fossa and an evident stretching of brainstem and CPA cranial nerves could be easily appreciated on MRI scans. The stretched nerves may be homolateral or contralateral to the site of the arachnoid cyst.

The mechanisms proposed to explain the atypical clinical presentation of the arachnoid cyst is the generation of a neurovascular conflict or an impairment of 'vasa nervorum' due to the stretching of the involved cranial nerve. In our patient, the degree of brainstem rotation, especially at the pons level, seems to be sufficient to cause significant mechanical stress to the left VIIIth nerve complex.

Conclusions

In adulthood, supratentorial arachnoid cysts usually present in a predictable manner, with headache, nausea, vomiting, visual disturbance with papilloedema; nevertheless, atypical presentation is also possible with unsteadiness, giddiness, ataxia and other symptoms attributable to a posterior fossa expansile mass. All these patterns of presentations seem to be caused by rotation or torsion of the brainstem with consequent stretching of the involved cranial nerves; the audiovestibular nerve for its course, position, and neural envelope seems to be the more sensitive to mechanical stress. Acute peripheral vertigo, aural fullness, and tinnitus resembling a Ménière's disease attack remain an exceptional mode of clinical onset.

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Address for correspondence:
Giuseppe Buongiorno, M.D.,
via Stellone 7,
10126 Torino,
Italy.

Fax: +39-011-6336642
E-mail: giuseppe.buongiorno@fastwebnet.it

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