Unusual association of diseases/symptoms

Asymptomatic supratentorial arachnoid cyst and symptomatic demyelination plaques: which one needs treatment?

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Summary

Intracranial arachnoid cysts are rare but the widespread use of brain imaging has disclosed many asymptomatic cysts. The authors report the case of a 37-year-old woman who presented with features suggestive of relapsing–remitting multiple sclerosis (MS). Her brain MRI revealed a moderately large arachnoid cyst in the right frontal area in addition to the periventricular plaques of MS. The cyst was an incidental imaging finding and the patient was asymptomatic prior to the development of MS.

BACKGROUND

Arachnoid cysts are rare and their symptomatology is highly variable. Our patient's moderately large and asymptomatic cyst was discovered because of the development of multiple sclerosis (MS). The former needs no treatment, while the latter requires interferon therapy. This case highlights the importance of a finding of coexistent neurological disease which may not need intervention.

CASE PRESENTATION

A 37-year-old woman presented with painful left-sided blurring of vision, which had been progressing over 1 week. She is married and the mother of two boys. She gave a history of right hand numbness a few months previously which had improved spontaneously over 1 week. In addition, her legs during the last year occasionally became very weak for 1 or 2 weeks and then recovered after a few weeks.

Figure 1 Axial T1-weighted brain MRI of the patient with contrast; some of the periventricular lesions have taken up gadolinium. There is a C-shaped enhancement of a large demyelination plaque in the right hemispheric deep white matter (inside the white circle), which is highly characteristic of multiple sclerosis. The arachnoid cyst did not take up the contrast medium.

Figure 2 Sagittal T2 fluid attenuated inversion recovery (FLAIR) brain MRI film of the patient; note the somewhat oval-shaped hypointense black region (red arrow) in the frontal area, which has been suppressed on this FLAIR film, indicating that the content of the cyst is cerebrospinal fluid. The periventricular demyelinating plaque lesions persisted and were not suppressed on the T2 FLAIR MRI.

Examination revealed left-sided reduced visual acuity to light perception, Marcus–Gunn pupil and normal fundus. All limbs were spastic and demonstrated exaggerated reflexes. Plantar reflexes were upgoing. A brain MRI (figures 1 and 2) showed multiple periventricular plaques, a few of which took up gadolinium, and a moderately large arachnoid cyst in the right frontal pole. The patient lives in a rural area without medical services, near the Iraq border.

INVESTIGATIONS

The authors conducted a battery of laboratory tests, which included antibodies panel and toxicology screen. All blood tests were normal. Her chest plain film and abdominal ultrasonography were unremarkable.

TREATMENT

The patient was given 1g of intravenous methylprednisolone daily for 3 days and a 2-week course of oral prednisolone was prescribed. Neurosurgical consultation revealed that the arachnoid cyst needed no surgical intervention.

OUTCOME AND FOLLOW-UP

The patient's left vision improved gradually over 1 month to finger counting. The patient was educated about her demyelinating disease and told that her intracranial arachnoid cyst was an incidental finding and needed no neurosurgery. Her expanded disability status score was 4, and the patient was prescribed interferon β injections.

DISCUSSION

An arachnoid cyst can develop when cerebrospinal fluid (CSF) collects between layers of the arachnoid matter of the meninges. Most of these cysts are said to be primary and arise from abnormal development of the brain meninges in utero. However, some cysts are acquired and result from loculation of CSF within layers of arachnoid mater because of inflammatory adhesions.

Some cysts enlarge gradually over time; Dyck and Gruskin¹ suggested that this enlargement might result from progressive secretion of CSF into the cysts by the lining cells, osmotic passage of CSF into the cyst, or entrapment of the CSF within the cyst by a ball-valve mechanism.

According to Pascual-Castroviejo *et al*,² 42% of arachnoid cysts are found in the supratentorial fossa; of these, 5% are located on the convexity of the brain (as in our patient). The cyst content never communicates with the lateral ventricles but may or may not communicate with the subarachnoid space. Meizner *et al*³ found that eight of their 32 cases of primary arachnoid cysts had additional central nervous system or other body anomalies, the commonest being cranial asymmetry.

Arachnoid cysts comprise approximately 1% of all intracranial space-occupying lesions. The widespread use of brain imaging has disclosed the presence of asymptomatic cysts. Most of these asymptomatic cysts were found in adults; in contrast, around 75% of cysts found in the paediatric age group were symptomatic. Persistent headache, epilepsy, focal deficits and cognitive impairment are the usual

manifestations in symptomatic patients with supratentorial arachnoid cysts. However, in their study, Spansdahl and Solheim⁴ found that there was great variation in the presenting symptoms, apparently without any relation to cyst localisation. They also concluded that individuals with arachnoid cysts had low employment status, decreased quality of life scores and prevalent symptoms of anxiety. Therefore, such cysts might not be entirely asymptomatic.

Although our patient admitted to having had pancephalic headache occasionally over the past few years, she first noticed body aches and stiffness 1 year ago; this complaint was investigated three times with thyroid function tests (the patient came from an area with endemic hypothyroidism), which were normal. This stiffness was punctuated by episodic weakness. However, she denied any muscle weakness, stiffness, sensory loss or seizures in the preceding years. The patient lives in a mountainous area on the Iraqi–Iranian border, which lacks medical services; this explains her delayed visit to our department in spite of having had recurrent neurological complaints over the past year.

The patient had developed relapsing–remitting MS, which was unrelated to the primary frontal arachnoid cyst. The pertinent literature mentions that surgical intervention should be considered in symptomatic cysts only. Immunomodulating therapy in relapsing MS is only moderately effective and associated with side effects. The long-term effect of these therapies is unknown. The use of immunomodulating therapy should follow shared decision-making by the patient and their neurologist after discussion of expectations and side effects.

The medical literature does not mention that arachnoid cysts are risk factors for the future development of MS. The authors think that, in our patient, both diseases simply coexisted with each other without any direct association.

Learning points

- ▶ Arachnoid cysts are often an incidental finding on cranial MRI.
- ▶ Arachnoid cysts are often asymptomatic and do not need neurosurgical intervention.
- ▶ Primary arachnoid cysts may be associated with central nervous system and other body malformations but they are not risk factors for multiple sclerosis.

Competing interests None.

Patient consent Obtained.

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