CASE REPORT

Middle fossa arachnoid cysts and inner ear symptoms: Are they related?
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Abstract
Background: Arachnoid cysts most frequently occur in the middle cranial fossa and when they are symptomatic, patients present with central nervous symptoms. Nevertheless, a large proportion of arachnoid cysts are incidentally diagnosed during neuroimaging in cases with nonspecific symptoms.

Report of cases: The cases of two males with middle cranial fossa arachnoid cysts with nonspecific inner ear symptoms were retrospectively reviewed. The first patient presented with mild headache, nausea, vertigo, unsteadiness, and tinnitus on the left ear while the second patient’s main complaint was left sided tinnitus. Both patients (initially managed for peripheral disorders) underwent a thorough clinical and electrophysiological evaluation. Because of the patients’ persistent clinical symptoms, and indications of CNS disorder in the first case, neuroimaging by brain MRI was performed revealing a middle cranial fossa arachnoid cyst in both patients.

Conclusion: Occasionally, patients with arachnoid cysts may present with mild, atypical or intermittent and irrelevant symptoms which can mislead diagnosis. Otorhinolaryngologists should be aware of the fact that atypical, recurrent or intermittent symptoms may masquerade a CNS disorder. Hippokratia 2014; 18 (2):168-171.

Keywords: arachnoid cyst, middle cranial fossa, atypical presenting symptoms, vertigo, hearing loss, tinnitus

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Introduction
Arachnoid cysts are rare cerebrospinal fluid (CSF) collections within the arachnoid membrane and subarachnoid space1. Histopathologic studies report their incidence ranging from 0.1 to 0.7% of all intracranial mass lesions2.

Most of the cases represent congenital lesions (primary arachnoid cysts) resulting from alterations in CSF flow in the early phase of subarachnoid space formation2. Congenital arachnoid cysts are usually found in Sylvian fissure and are classified into three groups, according Galassi, depending on their size and relation with Sylvian fissure3,4. Although some cysts remain stable, the majority become symptomatic in early childhood, with 60-90% of all patients with arachnoid cysts being children2,5,6. Symptoms of primary arachnoid cysts usually appear during infancy, but delayed presentation until adolescence has also been reported6. In children arachnoid cysts occur more frequently in males, while in the elderly they are equally distributed2,6.

Secondary arachnoid cysts may also be developed in metabolic diseases of the brain, after head trauma, subsequent to meningitis, brain tumor/ hemorrhage or iatrogenically following neurosurgical procedures or after over-rainage of CSF7. Although arachnoid cysts can occur at any site where arachnoid membrane exists, these lesions commonly can be found on the surface of the brain at the level of the main brain fissures. Approximately half of them are located in middle cranial fossa10.

Arachnoid cysts may be present with headache, seizures, increased intracranial pressure, hydrocephalus, ataxia, hemiparesis, focal neurological signs and behavioral changes7. Furthermore, patients may experience nonspecific symptoms such as confusion, sleep apnea, nausea, vomiting, tinnitus, hypoacusia, dizziness and unsteadiness7. Manifestation of these less typical symptoms, especially when they are not severe, reduces the possibility of early diagnosis and subsequent adequate management. Despite the possible presenting symptoms, a large proportion of arachnoid cysts are incidentally diagnosed during neuroimaging.

We report the cases of two adults with middle fossa arachnoid cysts referred to our department with nonspecific inner ear symptoms.

Case 1
A previously asymptomatic 56-year-old male was referred to the ENT Department of Chania General Hospital with the complaints of mild headache, nausea, vertigo, unsteadiness and tinnitus on the left ear. Patient reported his symptoms to be intermittent for the last four weeks. He was treated by his general practitioner with a five-day course of vestibular suppressants with no clinical improvement. Ear examination revealed normal appearance of tympanic membrane and middle ear. Neurotologic examination revealed a low-amplitude nystagmus, with Frenzel glasses, that was not seen during visual fixation. The nystag-
nus was conjugate and predominantly horizontal with the fast component beating to the right only at the right gaze position (1st degree nystagmus according to the Alexander’s law). There was a torsional component with the upper poles of the eyes beating towards the right ear. No focal neurological deficit was found, while Romberg’s test was positive and patient had wide-based gait, with slow cadence. Pure tone audiometry revealed mild sensorineural hearing loss on the left (Figure 1), while tympanometry was normal. Patient was treated with a second course of vestibular suppressants as well as steroids in a tapering manner. Nystagmus was the only symptom that subsided. Auditory Brainstem Response (ABR) showed no significant increase in wave V latency on the left and only a slight prolongation of the wave III latency on the left was recorded (3.97 ms) (Figure 1). Evaluation of the vestibular system was performed on a scheduled appointment after patient’s discharge, with video-oculography and video-nystagmography revealing pathologic smooth pursuit test and saccades (Figure 2).

Smooth pursuit eye movements were elicited by means of a projected yellow- colored circular target with a width of 10°, moving in a sinusoidal pattern in horizontal plane. Stimulus movement lasted 45 sec had an amplitude of 10°, a turn break of 0.0 sec and a velocity of 100°/sec. A projected yellow- colored circular target with 0.3 width, moving randomly in horizontal and vertical planes, was used eliciting saccade eye movements. Imaging was decided due to the intermittent patient’s symptoms, as well as, the findings of auditory brainstem response and video-oculography. Brain MRI revealed a large cystic lesion occupying the middle fossa causing ventricle asymmetry (Figure 3). The diagnosis was middle fossa arachnoid cyst (Galassi type III) probably of congenital origin, as the patient did not report history of CNS inflammation or any neuro-surgical procedure. Neurosurgeon’s consultation was asked and the patient was managed with oral acetazolamide (15 mg/Kg/day in three doses). Medication was discontinued 12 weeks later when all the patient’s symptoms subsided. The patient has been informed for the possible complications of his condition, as well as, for the need of follow-up and the several treatment modalities. A year after his admission, the patient is free of vestibular symptoms with unchanged hearing thresholds. Follow-up MRI revealed no enlargement of the arachnoid cyst.

Case 2

A 48-year-old male was admitted to the ENT department of Chania General Hospital with the main complaint of tinnitus on the left ear lasting for a week. Clinical examination revealed normal otoscopy and endoscopy of the upper respiratory system. He did not report history of imbalance and clinical evaluation of the vestibular system was uneventful. Pure tone audiogram revealed a symmetric high frequency hearing loss (Figure 4), while ABR was normal with an interaural latency difference (ILD) for wave V of 0.13 ms at stimulus intensity 90dBnHL. His tinnitus was tonal, with a pitch of 8KHz and an intensity of 9 dBnHL. The patient received
β-histine 24mg b.i.d. for two months with a reported aggravation of his symptoms. Brain MRI revealed an arachnoid cyst at the anterior part of the temporal lobe (Galassi type I) on the left (Figure 5). The patient did not report history of CNS inflammation or any neurosurgical procedure and has been followed-up by the neurosurgical department of the hospital with brain imaging at regular intervals. Two years after initial evaluation no enlargement of the arachnoid cyst has been noticed. He reports improvement of his tinnitus after tinnitus retraining therapy with sound generator.

Prevalent hypothesis for primary arachnoid cysts formation is arachnoid tearing after complete differentiation in 15th week of gestation or during separation between arachnoid membrane and dura mater. Although these lesions present symptoms during childhood, they may give symptoms later in adulthood, or remain asymptomatic. Symptoms might be caused by a space occupying effect with pressure on surrounding structures, a change in CSF dynamics, or dysgenesis of the brain. Late symptoms presentation is attributed to intracystic bleeding and subdural hematoma following head trauma, or to spontaneous cyst expansion. A wide range of symptoms at the initial patients’ evaluation have been reported which were seemingly unrelated to the location of the cyst. Thus, patients may present with atypical signs and symptoms such as sleep apnea, nausea, vomiting, tinnitus, unsteadiness, vertigo, hypoacusia, which initially may not attributed to central nervous system lesions and therefore adequate management is delayed.

Arachnoid cysts, as in our cases, have a predilection for males and the left temporal fossa. Our first patient treated initially as a subacute cochleovestibular disorder. Although his gait was characterized as ataxic, in the absence of any focal neurological deficits or cerebellar pathological signs, we concluded that it reflected an uncompensated vestibular disorder in which, the signals from inner ear were unable to reach brainstem and cerebellum for balance and gait adjustment. The pathologic findings of the vestibular system evaluation (abnormal smooth pursuit and saccades) and in ABR (prolonged absolute latency of wave III on the left) imposed brain imaging. Tunes et al suggest that dizziness and vertigo is the pressure effect of temporal arachnoid cysts on the vestibular cortex. In the central vestibular system secondary vestibular afferents relay signals from vestibular nuclei to the extraocular motor nuclei, the spinal cord, or the flocculus of the cerebellum. Many vestibular reflexes are controlled by processes that exist primarily within the brainstem. Tracing techniques have identified extensive connections between the vestibular nuclei and reticular formation, thalamus and cerebellum. Vestibular pathways appear to terminate in a unique cortical area. Recent studies in humans confirm the parietal and insular regions as the cortical location for processing vestibular information. Furthermore, a vestibular area has been identified in the temporal cortex, which could be linked to the perception of self-motion. The pursuit pathway starts at the retina and runs to the magnocellular portion of the lateral geniculate nucleus, the striate cortex, the dorsolateral pontine nucleus, cerebellum, vestibular nuclei, brainstem reticular formation and the oculomotor nuclei. Saccades are produced and controlled via the occipitoparietal cortex, the frontal lobes, the basal ganglia, the superior colliculus, the cerebellum and the brainstem. Central auditory afferent pathway includes cochlear nucleus, superior olivary complex, lateral lemniscus, inferior colliculus, medial geniculate body and auditory cortex. In the first patient electrophysiologic evaluation showed a delay of the signal transmission from the distal part of cochlear nerve to the superior olivary complex, probably due to the compression...
of the acoustic pathway structures along the brainstem.

The second patient’s main complaint was tinnitus, which showed aggravation during his completion of audiological and electrophysiological evaluation while he was receiving β-histine. He underwent brain MRI as part of our MRI evaluation protocol. β-histine has been reported for tinnitus treatment although, its effectiveness has not been established with prospective controlled studies. We prefer to prescribe a short course of β-histine in patients with tinnitus until their final evaluation, prior to other treatment modalities. We believe that a pressure effect following a spontaneous cyst enlargement might also be the reason of tinnitus, as tinnitus is an active physical process occurring in multiple neural substrates in response to peripheral or central stimulus.

Spansdahl and Solheim in their quality of life study in adult patients with primary intracranial arachnoid cysts, concluded that patients with atypical symptoms have a low employment status, decreased quality of life scores and prevalent symptoms of anxiety. Furthermore, clinical outcome for these patients seems to be worse than those with plausible symptoms because of late referral and delayed diagnosis.

With the advent of imaging studies, there has been an increased incidence of detection of incidental asymptomatic or with atypical symptoms arachnoid cysts. On CT scan, arachnoid cysts are well circumscribed, with an imperceptible wall displacing adjacent structures. When large, and over time, they can exert a remodeling effect on the bone. On MRI, arachnoid cysts follow CSF on all sequences, including FLAIR and DWI. This enables differential diagnosis from epidermoid cysts. As their wall is very thin it only occasionally can be seen, and their presence is implied by displacement of surrounding structures. As there is no solid component, no enhancement can be identified. Phase contrast imaging can also be employed not only to determine if the cyst communicates with the subarachnoid space, but also to identify the location of this communication. Differential diagnosis of arachnoid cysts includes enlarged CSF space (e.g. mega cisterna magna), epidermoid cyst, subdural hygroma, chronic subdural haemorrhage, pilocytic astrocytoma, haemangioblastoma, non-neoplastic cystic lesions, neurocysticercosis.

Asymptomatic patients are not operated on, while symptomatic patients are candidates for surgical intervention via several methods, including cystoperitoneal shunt placement, craniotomy, endoscopic fenestration and stereotactic aspiration.

Literature review shows that it is often difficult to interpret the relationship between inner ear symptoms and intracranial arachnoid cysts. In fact there is a debate whether arachnoid cysts are associated with these symptoms in most cases. We suggest that, inner ear symptoms might be attributed to middle fossa arachnoid cysts. Otorhinolaryngologists should be aware of the fact that atypical, recurrent or intermittent symptoms may masquerade a CNS disorder. Complete clinical and electrophysiological examination is needed and any pathological finding should be carefully evaluated for a prompt diagnosis.

Conflict of interest
The authors declare no conflict of interest.

References