Gusher phenomenon and Ménière disease. An enigmatic Case report

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Abstract
During stapedotomies or cochlear implant placement, cerebrospinal fluid gusher was described. The recognized causes result from abnormal communication between perilymphatic space and subarachnoid space. Our clinical case describes a patient with Ménière disease that presented a gusher phenomenon during surgery:- A dubious and controversial case report.

Introduction
The gusher phenomenon happens more frequently during stapes surgery and cochlear implant placement. Violent perilymph leakage starts during platinotomy or electrode placement and must be plugged in order not to cause sensorineural deafness. The stapes surgery is not feasible whereas, the positioning of cochlear implants is possible after the cleansing of cochleostomy. Moreover, other cases of uncontrolled perilymph leakage are described in the literature after temporal bone fracture. This phenomenon is due to abnormal communication between perilymphatic and subarachnoid spaces. The connection is produced by anatomical alterations such as internal auditory canal bone fistula or the cochlear and vestibular aqueduct enlargement. Mondini cochlear malformation is a cochlear gyrus reduction in the osseous and membranous labyrinth; the cochlea does not finish its development during the seventh week of gestation. Cochlea dysplasia is often associated with fistulas between a labyrinth and the subarachnoid spaces (modiolus fistula).

The cochlear aqueduct allows communication between the cochlear perilymphatic space and the cerebellar subarachnoid fossa. This structure plays a role in perilymphatic, endolymphatic, and cerebrospinal fluid flow. The fluid flow rate is a function of the fourth power of duct radius, therefore a minimal change of cochlear aqueduct radius is responsible for a change in the inner ear hydrodynamics. To avoid the high risk of hearing loss due to cerebrospinal fluid (CSF) gusher, a proper diagnostic procedure is required before this type of surgery.

Case presentation
The 81-year-old patient reports vestibular symptoms for about 30 years with objective vertigo lasting about an hour with fullness and tinnitus referable to the left ear.

Serial audiometric examinations showed a fluctuating sensorineural hearing loss in the left ear, ranging from low to medium frequencies (Fig. 1).

Tympanometry produced a type A graft and absent stapedius reflexes. In two different hospitals, Ménière disease (MD) diagnosis was made. Cranial CT excluded malformations of the inner ear: enlargement cochlear aqueduct (ECA), enlargement vestibular aqueduct (EVA), Mondini Malformation, third-window lesion, ear semicircular canal fistula (Fig. 2). Also in our hospital, after excluding other pathologies with vestibular symptomatology, a diagnosis of
Definite Meniere disease Meniere disease of the left ear was confirmed following the Classification Committee of the Bárány Society.\textsuperscript{10,11}

Every ten years, audiometric examinations showed severe left pantonal sensorineural hearing loss (Fig.1). The patient described a pause in vertigo crisis episodes from 2001 until January 2018, when a more severe crisis occurred, with objective vertigo lasting 6-7 hours with nausea and vomit. (betahistine, diuretics, dexamethasone) failed to treat vertigo. Left transcanal labyrinthectomy has been performed considering untreatable vertigos, patient’s age, and severe hearing loss.\textsuperscript{12}

During the intervention, the authors proceeded to tympanic cavity exploration. The middle ear appeared free from inflammatory disease and perilymphatic fistulas of the windows; therefore, with a microscopic vision, two windows ossification were identified. During the opening of the oval window, unpredictable high-pressure liquid leakage occurred with a diagnosis of the perilymphatic gusher. The following step was to put a fragment of the temporal muscle and temporal fascia on the oval window and the round window until we verified the absence of perilymph.

**Discussion**

The perilymphatic gusher was described in stapes surgery and cochlear implant placement. Otosclerosis results in bilateral mixed or conductive hearing loss with absent stapedial reflexes; only rarely high-resolution CT scan or an MRI is required for the inner ear studies, therefore it is difficult to suspect possible gusher events.

The inner ear should be studied with radiology before surgery to avoid the risk of hearing loss.

A cranial CT scan can highlight cochlear and vestibular aqueduct enlargement, the Mondini malformation, and other forms of inner ear hypoplasia, but not allow evaluate inner ear canal fistulas.\textsuperscript{13} Several studies also argue that an enlarged vestibular aqueduct is not necessarily associated with the gusher phenomenon.\textsuperscript{14} However, there seems to be a more significant correlation with the enlargement of the cochlear aqueduct.\textsuperscript{13,15}

Other studies show that an oblique plane CT scan can diagnose bone dehiscence between the basal gyrus of the cochlea and the inner ear canal.\textsuperscript{16,4} Preoperative CT scans does not necessarily investigate subtle inner ear malformations.

A retrospective study on preoperative CTs revealed that this exam could be associated with false negative for this pathology. A correct diagnosis is essential to avoid the gusher phenomenon, but a CT scan does not always help the surgeon. This study is the first reported case of a perilymphatic gusher in a patient suffering from vertigo. CSF leakage has never been described in a patient with MD.
Among Meniere's disease therapies, No window opening procedures are reported. Labyrinthectomy can be used as the last step of MD treatment, but no cases of CSF gusher have been described during transcanal labyrinthectomy. More data is needed in the literature to answer these doubts.

Based on the data collected, the authors formulate the following hypotheses to explain the clinical case:

- Prominent endolymphatic hydrops could be the cause of a gusher phenomenon
- MD in vertigo absence, if unilateral, can mimic otosclerosis.
- The patient could be affected by MD and an unrecognized inner ear malformation.
- The MD diagnosis could be incorrect because of a malformation of the inner ear that could create a diagnostic picture similar to MD.

**Conclusion**

Internal ear abnormalities can cause no-standard communication between perilymphatic and subarachnoid space; these anomalies may remain unknown. Can Meniere's disease be considered an endolymphatic hypertension?

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**References**