CASE REPORT

Late pneumolabyrinth after undiagnosed post-traumatic perilymphatic fistula. Case report illustrating the importance of systematic emergency management

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Introduction
Temporal bone fracture is a common complication of high-energy cranial trauma. Labyrinth involvement is rare, but there is a risk of perilymphatic rupture that is often underestimated on initial clinical examination due to the predominance of neurological and/or somatic symptoms. Based on an illustrative case report and a review of the literature, we describe a systematic management protocol in cranial trauma with suspected temporal bone fracture to prevent late complications.

Case report
A 34-year-old man was admitted to the emergency department following craniofacial trauma by direct shock, leading
to loss of consciousness, sustained in a fight. The patient was under the influence of alcohol at initial examination, and complained of headache, loss of memory of the incident, and multiple pain, notably in the lower jaw. As he mentioned no cochleovestibular symptoms, no ENT opinion was sought. Clinical, and notably neurological, examination in the emergency setting was normal, without cochleovestibular signs. Emergency brain CT found slight right meningeal hemorrhage and fracture of the right mandibular angle. The tympanomastoid cavities were free, with no effusion suggestive of temporal bone fracture (Fig. 1). The patient was admitted to post-emergency surveillance. During the following night, he experienced left auditory discomfort, with hypoacusis and a "blocked ear" sensation which led him to attempt numerous Valsalva-type hyperpressure maneuvers; he then reported rapidly progressing onset of vertigo and left-ear tinnitus with evident hearing-loss. An emergency ENT opinion was sought only the next morning (it is to be borne in mind that the patient, under the influence of alcohol, was difficult to manage and uncooperative).

Clinical examination found left hemotympanum, ipsilateral hypoacusis and right lateralization on Weber test. The patient showed nausea, left segmentary deviation and right horizontal-rotational nystagmus. An audiogram and tympanometry found left-sided cophosis and loss of left stapedial reflex on ipsilateral stimulation.

Emergency temporal bone CT with millimeter-scale slices found a longitudinal left submeatal fracture and pneumolabyrinth. There was also mastoid cell filling, effusion at the vestibular window with an aspect of stapes footplate fracture, and continuity of the tegmen tympani (Figs. 2 and 3). Medically, the patient was given symptomatic treatment (anti-vertigo, anti-nausea), inner-ear protection treatment (i.v. corticosteroids, vasodilators) and prophylactic antibiotic therapy (amoxicillin plus clavulanic acid) as well as a vaccination (anti-pneumococcal, anti-haemophilus influenzae, anti-meningococcal). Emergency surgical exploration was undertaken in view of a strong suspicion of perilymphatic fistula. Examination of the retrotympanum found perilymphatic liquid leaking from the vestibular window via a footplate fracture and from the cochlear window (Fig. 4A). The two fistulae were treated by apposition of temporal muscle and biologic glue, conserving the stapes, and with a perichondrial fragment interposed between the two stapedial crura, respectively. There was also a fracture line at the tegmen, with cerebrospinal fluid (CSF) leakage (Fig. 4B), which was sealed by temporal muscle and aponeurosis apposition reinforced with glued bone powder.

Immediate postoperative course featured progressive improvement in vertigo and tinnitus, but not in hearing. Retrospective re-reading of the initial CT data, using a lower bone-density threshold than usual, revealed a slight pneunolabyrinth (Fig. 5).

One month postoperatively, the patient showed non-disabling instability; leftward index finger deviation without spontaneous nystagmus persisted, with deafness. Vestibular rehabilitation was re-initiated, leading to complete resolution of the instability by 6 months' follow-up.

**Discussion**

Temporal bone fracture is always a sign of high energy cranial trauma. Emergency management includes painstaking interview to explore for not only neurological but also otological signs. Apart from left hearing loss, the symptoms suggestive of perilymphatic fistula are positional vertigo aggravated by closed-glotis pressure effort or tragal pressure inducing nystagmus; these maneuvers can be performed in the emergency setting, even if the patient is poorly compliant or in a state of altered consciousness. Cranial trauma with temporal bone fracture is frequently associated with no inner ear involvement; cranial trauma without temporal bone fracture but with vestibular or cochlear involvement, as in the present case, on the other hand, is much rarer [1]. Even when there are no manifest otologic signs, otologic involvement (labyrinthine concussion, fistula or hemorrhage) should always be suspected in cranial trauma in the broadest sense, and be specifically explored. Cochleovestibular signs are therefore among those to be systematically looked for in analyzing cranial nerve-pair damage. These symptoms may well have been underestimated in the present case, as the patient was under the influence of alcohol and presented with a mandibular fracture that was initially managed without thought of any neurosurgical emergency. It should be stressed that a reassuring initial clinical examination does not rule out temporal bone fracture with complications: an ENT opinion is
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Figure 2  Pre-operative CT-scan. Millimeter-scale axial slices centered on the bone windows. Tympanomastoid effusion; anterior and posterior pneumolabyrinth (A and B, arrow-heads) and stapes footplate fracture (A, arrow).

Figure 3  Pre-operative CT-scan. Millimeter-scale coronal slices centered on the bone windows; effusion predominating in the posterior labyrinth (B, arrow); anterior and posterior pneumolabyrinth (A, arrow-head).

Figure 4  Peroperative views. A. Oto-endoscopic view (30° angle) of the oval window region: anterior branch fracture (arrow-head) and air bubbles between the two stapedial crus, inward of the footplate (arrow), indicating pneumolabyrinth (i: incus; f: facial nerve). B. Microscopic view: squamous temporal fracture extending to the tegmen tympani (arrows).

Figure 5  Initial brain CT. Enlargement centered on temporal bones. A and B. The mastoid cells and tympanum are free of effusion. C. Enlargement, of poor quality, due to the scan resolution, nevertheless shows a millimeter-scale bubble on the deep side of the footplate (arrow) (in a superior position due to the properties of air in a liquid interface).
mandatory. Likewise — and this is the take-home message of the present case — labyrinthine concussion should be systematically suspected in cranial trauma. If, in the present case, the initial or the late cochleovestibular symptoms had been noted on admission to the emergency department, the patient could have been warned against hyperpressure effort, and his deafness might have been avoided. ENT examination would not have enabled early diagnosis and surgical treatment of the perilymphatic fistula: the patient was asymptomatic and the initial CT report did not point to it; but adapted management and advice could have been provided.

Temporal bone fractures are classified, according to the relation of the fracture line to the plane of the temporal bone, as longitudinal, transverse or complex [2]. An alternative radioclinical classification according to inner-ear involvement distinguishes extra- and intra-labyrinthine temporal bone fracture [3], providing a better description of the close relation between temporal bone fracture and the intra-temporal functional and/or vital structures.

Absence of temporal bone fracture on initial brain CT (generally intended as a rough and ready overview) does not rule out vestibular or cochlear window or ossicular chain involvement. In the present case, careful analysis of the bone-window CT slices could have detected a slight pneumolabyrinth, signalling perilymphatic space rupture, and raised a doubt about possible tegmen rupture (Fig. 5). We recommend systematic examination of the cochleovestibular apparatus in all cases of cranial trauma, by meticulous examination of the labyrinthine apparatus on the initial CT images. Ideally, brain CT should be completed by high-resolution millimeter-scale slices centered on the temporal bone [4]. Brain CT should be associated to bone-window slices that could highlight such a lesion. Millimeter-scale CT provides precise lesion analysis, explaining clinical signs and guiding treatment. MRI, on the other hand, is unsuited to the emergency setting, but is an indispensable complementary examination for studying the labyrinth and the acoustic-facial bundle when CT has detected abnormality. In some cases of diagnostic difficulty in second-line management, lowering the bone-density threshold in the neighborhood of the air (thresholding technique) can provide further information [5]. Thresholding associated to virtual endoscopy can detect labyrinthine fistulae of less than 0.3 mm, which usually go unnoticed on classic axial slices. The radiological signs of labyrinth fracture are pneumolabyrinth, otic capsule fracture and stapedial fracture or dislocation. Pneumolabyrinth or pneumocochlea is, by definition, the presence of air in the cochlea or vestibule, indicating a pathologic communication between the inner ear and tympanomastoid cavities. Pneumolabyrinth is thus characteristic or indeed pathognomic with respect to perilymphatic fistula [6]. Air in the inner ear is not physiologically normal: pneumolabyrinth is not uncommon in complex, transverse or intralabyrinthine fractures, but is often transitory; detection depends mainly on examination performed soon enough after the trauma. The impact of air in the inner ear is unclear. Perception deafness was experimentally induced in animals by introducing air bubbles into the labyrinth [7], where they disturbed electrical transmission along the basement membrane. Degree of hearing loss was also shown to depend on the position of the air within the inner ear: air inside the vestibular ramp had a greater and often irreversible impact on hearing, whereas air in the tympanic ramp induced milder loss which was often reversible once the air had been reabsorbed [6–9].

As seen in the present case, there may be asymptomatic perilymphatic fistulae in both windows that can decompensate under hyperpressure effort. In the present case, the communication was due to stapedial footplate fracture inducing perilymphatic fistula. The fistulae may be silent if they are not or only slightly permeable, but at great risk of decompensation if hyperpressure is applied voluntarily (Valsalva maneuver) or involuntarily (nose-blowing). In the present case, repeated Valsalva effort exacerbated the fistula and pneumolabyrinth, inducing deafness. Early diagnosis would have allowed the patient to be preventedly warned against this complication.

Management options comprise observation in hospital under medical treatment (corticosteroids, rest, anti-vertigo treatment and vasodilators), or surgery. Temporal bone fracture is considered as a serious trauma to the middle skull base; victims should be systematically vaccinated against pneumococcus, according to French Neurology and Neurosurgery Society guidelines; anti-meningococcal or anti-haemophilus influenzae vaccination is not recommended. The place of prophylactic antibiotic therapy in skull-base fracture, with or without CSF leakage is controversial [10]: Brodie and Thompson [11], in a series of 122 patients with temporal bone fracture and CSF leakage, arrived at no conclusive recommendations, whereas, in a separate meta-analysis, Brodie [12] demonstrated a significant increase in onset of meningitis in patients with CSF leakage who had not received prophylactic antibiotherapy. The French Neurology and Neurosurgery Society has no concrete recommendations on prophylactic antibiotherapy trauma of the anterior skull-base.

Surgery is recommended in fluctuating perceptual hearing loss, progressive hearing loss or persistent vertigo [13,14]. This option was taken in the present case in line with these recommendations. The types of lesion found comprise footplate fracture with vestibular window fistula, annular ligament detachment or stapes dislocation or cochlear window membrane rupture. Pneumolabyrinth location may help locate the leakage site; especially when the vestibule is primarily involved, oval window fistula is highly probable. (Fig. 3B).

In case of doubt, there are “easy” ways of helping detect fistula peroperatively: tilting, hyperventilation or jugular compression; such exploration, however, presupposes general anesthesia. One of the most reliable indirect signs is visualization of reflected microscope light in the moving liquid [15].

Surgeons have various means of ensuring tight fistula closure: apposition of tragal or conchal perichondrium, fascia temporalis or fatty fragments or using biologic glue to improve repair stability. The question of stapes conservation may arise in some rare cases, depending on the type of lesion: fracture, dislocation, annular ligament tear, or footplate luxation into the vestibule.

In case of fracture, subluxation or annular ligament tear, it is sometimes recommended not to remove the stapes, so as not to worsen auditory status; this attitude is especially
justified when surgical exploration is late and the stapes has luxated deeply into the vestibule, with increased risk of traction on footplate adherence to the membranous labyrinth [14].

In case of stapedial luxation into the vestibular window, treatment depends on depth [16]: when deep, leakage is hard to ensure against and it is preferable to remove the stapes so as to avoid fibrous adherence with associated maculae liable to induce late cochleovestibular impairment [14,16,17].

According to the literature [14], prognosis in pneumolabyrinth is good for vestibular symptoms, but not for recovery of hearing. The review by Tsubota et al. [14], on the other hand, showed that the shorter the interval to surgery, the better the functional prognosis, with optimally conserved hearing for trauma-to-surgery intervals of less than 10 days.

Conclusion

Cranial trauma is always serious and may lead to inner-ear damage, even when initial examination finds no cochleovestibular signs. Only specialized cochleovestibular examination can detect minimal signs and thereby prevent exacerbation of cochlear or vestibular compromise. Initial CT examination in skull base trauma should include high-resolution slices centered on the inner-ear, with careful interpretation by a craniofacial trauma specialist. When cochlear fracture or fistula is suspected, emergency surgical exploration is recommended after high-resolution CT-scan, to prevent onset of late deafness.

Disclosure of interest

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

References