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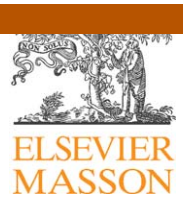

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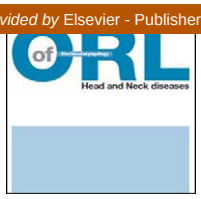

CLINICAL CASE TO THE EXPERTS

Sudden deafness and neurinoma☆

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Case

A 32-year-old air hostess, with a passion for music, consulted the day after a third episode of so-called ''sudden'' deafness in the left ear. The other two episodes, 1 and 3 years previously, had both been managed by corticotherapy, without hospital admission, resolving completely in a matter of days. There had been no associated vestibular system symptoms. Given the rapid resolution, the patient's physician had not undertaken further investigation. The patient had no particular history, and no brothers or sisters; both parents had died in a road accident many years before. Audiometry, performed at consultation, showed ascending (i.e., predominantly low-frequency: 35 dB at 500 Hz, 25 dB at 1 kHz, and 15 dB at 2 kHz) perceptual hearing loss. Clinical and paraclinical vestibular examination, including caloric test and vestibular evoked myogenic potentials (VEMP), was normal, as was tympanic membrane examination.

Questions

Question no. 1: what other examinations would you seek for this patient, who is worried by this third episode of sudden deafness? What diagnoses might you consider?

A. Chays

Before seeking further examinations, I would take up the interview again. I'm worried about these three episodes of left unilateral deafness, for which no diagnosis has as yet been determined, and am rather astonished that such unilateral symptomatology has not yet led to a more ''aggressive'' diagnostic approach, especially since the patient has a job that is at-risk for the ear, and is moreover a musician.

So I would try to clarify the circumstances of onset and any accompanying semiology, however transitory or nearly imperceptible.

After the interview, I would undertake complete clinical assessment, looking for the slightest neurological sign of urgency and the slightest vestibular symptoms of vestibulocochlear pathology. I would focus on the nasopharynx and its ganglion chains. I would also check the pulse regularity and blood pressure.

If the clinical assessment proves normal, I would prescribe an MRI scan focusing on the inner ear canals, the pontocerebellar angles, and the posterior fossa, particularly on the left. I would ask for a neurological brain exploration

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if retrocochlear pathway exploration fails to provide a solution.
I would also prescribe blood tests: CBC, ESR, CRP.
I would complete tonal with vocal audiometry.

To the patient herself, I would simply say that a pathology ''along the auditory nerve'' is the first possible diagnosis to try to rule out. I would explain that if results are normal, as expected, there will be further diagnostic examinations.

C. Dubreuil
This young patient shows totally regressive episodes of deafness, without vestibular involvement on clinical or instrumental exploration: there is no functional impairment of the inferior (normal sonomotor potentials) or superior (normal caloric test) vestibular nerves.

This is thus an isolated pathology of the cochlear pathways and/or cochlea itself.

Diagnosis could be refined by studying otoacoustic emissions (unfortunately, not covered by the French national health insurance system) and brainstem auditory evoked potentials (BAEP), so as to distinguish between purely cochlear and retrocochlear involvement: in the latter case, OAEs are normal and BAEPs abnormal.

The shape of the hearing curve is interesting: low-frequency loss with high frequencies conserved? This is suggestive of purely cochlear hydrops, without vestibular involvement or Ménière type syndrome, which would explain the total regression of the previous episodes and exclude any thrombotic phenomenon in this young patient (e.g., in case of estroprogestatives). Stress is usually the determining factor in these situations, which is to be kept in mind should MRI prove normal.

I would in any case ask for T1—T2 weighted MRI, possibly gadolinium-enhanced to shed greater light, studying not only the internal auditory canal but also the cochlea and pontocerebellar angle.

One possible diagnosis not to overlook is intracochlear neurinoma, with purely auditory involvement, the vestibule, both superior and inferior, being conserved.

F.-M. Vaneecloo
The audiometric curve essentially suggests a pressure-related etiology (labyrinthine hydrops). The patient should be asked whether she has ever experienced vertigo, even in the mildest form, tinnitus or fluctuating hearing loss.

As for paraclinical examinations, I would prescribe gadolinium MRI to study the inner ear, inner auditory canals and pontocerebellar angle, as well as some endocranial slices.

I would also ask for a basic biological assessment, specifically focusing on CRP, ESR, thyroid insufficiency and specific serology — the latter two to rule out thyroid insufficiency or neurolabyrinthine infection, however rare this may be.

Finally, I would probably schedule a tympanic ECOG, useful for diagnosing hydrops.

J. Magnan
I would ask for the following examinations: auditory evoked potentials and endocranial MRI, to decide between tumor (positive MRI) and cochlear hydrops (negative MRI).

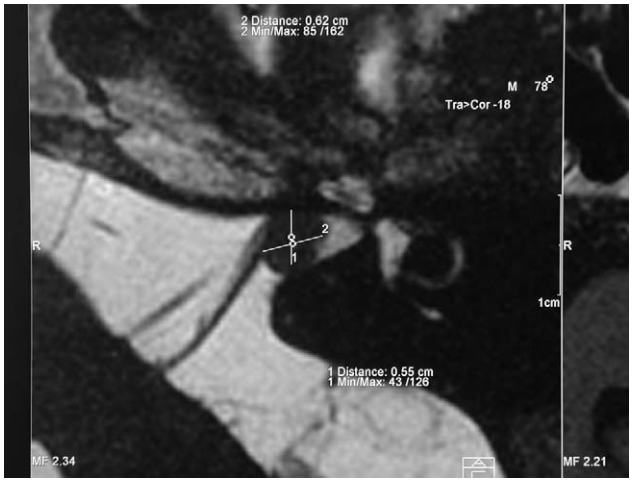

Figure 1 Axial T2-weighted MRI showing a small tumor of the left internal auditory meatus highly suggestive of a vestibular schwannoma.

Question no. 2: this is the MRI image of the fundus of the internal auditory canal. The suggested diagnosis is, of course, ''acoustic neurinoma''. What do you make of the clinical history? How do you interpret exam findings?

A. Chays
The MRI image shows a ''formation'' in the fundus of the internal auditory canal (IAC), not filling the far end (Fig. 1). It is basically spherical, perfectly contoured, a few millimeters in diameter and exerting no mechanical effect on the adjacent nerves: there is a thin layer of cerebrospinal fluid (CSF) between it and the IAC walls, and the two nerves that can be made out in the image are free in the CSF, although this would need confirming on a sagittal slice.

Other images should also be analyzed: Fat-Sat, to rule out any lipomatous element, and injection images to assess lesion contrast-medium uptake.

Finally, certain discrepancies between clinical and complementary exam results should be borne in mind:

- First episode of deafness 3 years ago, first recurrence 1 year ago. The lesion remains small and mechanically inactive, and may not in fact explain these episodes.
- The audiometry findings are not those expected for a lateral IAC lesion reaching the fundus, which would induce pure perceptual hearing loss predominating on high frequencies rather than an ascending-type curve.
- Vestibular examination, and notably VEMP exploration of the inferior vestibular nerve, was normal, which is unusual in acoustic neurinoma.

C. Dubreuil
The MRI confirms diagnosis of a rounded lesion in the fundus of the IAC. It is very likely a schwannoma, most probably originating as a schwannoma of the cochlear nerve: it is separate from the cochlear recess, and thus cannot have originated in the cochlea.

All in all, this is a rare location (< 5%) for a schwannoma of the cochlear nerve at the end of the cochlear recess, with-

out clinical or electrophysiological impact on the vestibular system.

F.-M. Vaneecloo
There is clearly IAC pathology. The entire MRI needs careful examination, especially to see whether this tumor-like image takes up contrast medium. There does not seem to be extension to the far end of the IAC. It would be interesting to know what light BAEPs could shed.

J. Magnan
The likely diagnoses are acoustic neurinoma, vestibular schwannoma or cochlear schwannoma. Detailed study of serial images in the various planes should determine the origin, not that this greatly affects treatment.

The clinical history of ''sudden deafness'' episodes at long intervals, with a very small canal lesion, is unusual. In neurinoma, however, the correlation between audiometric impairment and lesion size defies even the most expert assessment.

Question no. 3: more precisely, does the MRI image of IAC schwannoma seem to you to explain the recurrent nature of this ''sudden'' deafness and its ascendant aspect? If so, what mechanisms would you suggest?

A. Chays
The question itself seems to be saying that we are now virtually sure that the patient has an IAC schwannoma.

In point of fact, however, this image hardly explains the recurrent nature of the three episodes or the ascendant audiometric curve.

One might at a pinch argue that the lesion grew rapidly, with a slight hemorrhage, compressing the auditory nerve, and then recovered: but that would have to have happened twice! There would have to have been inflammation around the lesion at least three times; and the low frequency hearing loss would be a matter of ''tonotopic dissociation'' of the auditory nerve fibers.

I am not happy with any of these explanations; finding no obvious mechanism, I keep an open mind, awaiting further questions.

C. Dubreuil
The mechanism of this sudden deafness is vascular. See: JB Charrier, P Tran Ba Huy, Surdités brusques idiopathiques. Ann Otolaryngol Chir Cervicofac 2005;122(1):3—17.

Sudden deafness types A and B (pressure-related and horizontal) have the best prognosis of recovery. They are probably due to transitory ischemia or vasospasm of the labyrinthine and/or cochlear artery — from which it is a short step to consider Ménière-type syndromes to be related to vasospasm and its cochlear repercussions.

Also, complete auditory recovery may indicate a tumor of the pontocerebellar angle; in any unilateral perceptual hearing loss of sudden onset, whether or not progressive/regressive, and with or without associated tinnitus, vertigo or instability, one examination is mandatory: MRI, especially as the limitations of electrophysiological exploration are well-known.

T. Mom and P. Avan (Ischémie cochléaire des données fondamentales aux espoirs cliniques. Ann Otolaryngol Chir Cervicfac 2008;125:301—8) have demonstrated that:

- the cochlea successfully resists ischemia of less than 10 min;
- in incomplete ischemia or vasospasm, recovery may occur even after 7 min of ischemia;
- in neurinoma, sudden deafness is due to reversible cochlear artery ischemia; and
- several cases of sudden deafness have been reported secondarily to vertebrobasilar insufficiency.

Such cochlear ischemia is also clearly seen in neurinoma surgery, by means of acoustic distortion products.

F.-M. Vaneecloo
I have no exact explanation for the recurrence of sudden deafness in relation to acoustic neurinoma. Several hypotheses have been put forward, including the vascular hypothesis, recognized by the possibility of pure endocochlear perceptual hearing loss associated with acoustic neurinoma.

Impaired inner ear hydraulics could also very probably account for the facts.

And there may also be extension of the neurinoma within the cochlea — although, as far as I know, medical imaging always settles this (contrast medium uptake by the cochlea).

At all events, the problem of sudden deafness is prognostically worrying, and to the best of my knowledge hearing cannot be conserved in such cases.

J. Magnan
The pathogenesis of recurrent sudden low-frequency hearing loss remains hypothetical.

On the one hand, it is not sure that there is any direct link between the episodes of so-called ''sudden'' deafness and the tiny neurinoma, which may just be an associated chance discovery — although one pathology may be thought to be enough for the patient.

On the other hand, the patient's job subjects her to variations in pressure. Have these normally asymptomatic ''dysbaric events'' become symptomatic in an ear put at risk by an auditory nerve tumor?

Finally, ischemia induced by auditory nerve or cochlear tumor is the most comprehensible and plausible hypothesis.

Question no. 4: the patient cannot tolerate the idea of having a tumor ''in the brain'', and wants treatment. What do you suggest?

A. Chays
If the patient cannot tolerate the idea of a ''tumor in the brain'', I would explain that:

- simple surveillance would still leave the tumor;
- so would radiotherapy.

That leaves surgery, which is also the only way to satisfy the patient's wish and to clear up the histology of the lesion and remove it completely.

I would, however, explain how unlikely it is that she would keep her hearing, especially as cochlear symptomatology has been flagrant in this case. Bearing in mind that she is a musician, I would underline this point.

I favor a retrosigmoid approach.

C. Dubreuil

It is easy to explain that this is a microtumor and that it isn't in her brain but in the pars petrosa, and that hundreds of patients have such a tumor and that, given its size and especially its location and the slow speed of its evolution, it is far from dangerous. On the contrary, in the present context, we can be sure that it is benign.

Giving reassurance is part of our job, and this case is psychologically straightforward to manage.

Finally, we have to explain to her that in her case surveillance is the attitude of choice, whatever she may think. Any surgical or radiation treatment would to my mind be absurd and moreover, for this music lover and air hostess, would mean risking everything — music and job — just because of a more or less severe deterioration in her hearing which has so far been fairly stable.

Finally, imaging and ENT examinations make surveillance remarkably simple to do, noninvasive and generally well adhered to.

F.-M. Vaneecloo

The patient is young, with more than 50 years' life expectancy before her.

So I certainly wouldn't recommend treatment by Gamma Knife.

I don't think we have enough experience to know whether this treatment is really without danger over the long term.

This is a small tumor, and surveillance is certainly feasible.

But the patient wants to be operated on. We need to know exactly why, and to explain the objectives and risks of surgery: notably, postoperative facial palsy.

In such a case, where the tumor is filling the lateral part of the IAC, I doubt that hearing can be conserved, especially as the patient has had several episodes of sudden deafness. The facial nerve must be protected at all costs, with translabyrinthine exeresis, causing definitive total deafness, to be borne in mind in indicating surgery.

In such a case, I would at least wait a few more months, to see what the patient finally decides; meantime, a surveillance control MRI scan could be performed.

J. Magnan

With such a small, strictly intracanal volume, surgery or radiotherapy cannot be indicated without first explaining the auditory risk, which in the case of flight staff is going to mean a change of posts.

So first of all, surveillance MRI, which has the double advantage of precisely confirming both diagnosis and the evolution of hearing and of the tumor.

Informing the patient that surgery can be avoided (as 30 to 40% of such tumors are nonevolutive) without endangering hearing is usually enough to reassure and persuade. The treatment should not be more aggressive than the tumor image.

Question no. 5: following this consultation, the patient opted for MRI monitoring. One year later, she was readmitted in emergency for aggravated hearing loss without signs of vertigo. Audiometry showed cophosis and left areflexia. MRI revealed a moderate increase in the size of the tumor, which was still within the IAC, which it filled completely. What is your attitude?

A. Chays

We are more or less back to where we were in question no. 4, with an extra argument in favor of surgery: cophosis.

I confirm my previous recommendation: surgical exeresis.

I have always been suspicious of the rather curious clinical aspect in this case: episodes of ''sudden deafness'', with unusual audiometric recovery for a schwannoma, rapid tumor growth, an aggressive lesion rapidly inducing cophosis, and discrepancy between clinical and radiological aspects. Before operating, I would ask for electrophysiological assessment of the facial nerve.

C. Dubreuil

The spontaneous evolution of the neurinoma has not been life threatening.

On the other hand, at 1 year's evolution under surveillance, the tumor may grow (in 15% of cases), and become more symptomatic or not. In the present case, the neurinoma has become slightly bigger and the ear entirely cophotic. The patient has won 1 year of hearing but could have won more: it's the way the lesion chooses to evolve that decides. Since evolution has led to cophosis, there is now nothing more to lose: the patient's youth indicates either surveillance (but why wait, since the neurinoma seems to be aggressive with respect to the cochleovestibular nerve, just to make surgery more complicated?) or surgery: the cophosis and invasion of the fundus of the IAC require a translabyrinthine approach, which can be performed rapidly with minimal risk to the facial nerve (1% rate of palsy). The patient will need to change posts at work.

F.-M. Vaneecloo

No Gamma Knife at her age. For this evolutive tumor, surgery with a translabyrinthine approach, if the patient has made up her mind.

J. Magnan

The natural evolution has simplified the issue.

This young patient will have to be operated on: sequelae are not to be feared, as she is already cophotic. Scheduling surgery and selecting the approach are matters of personal preference for the surgeon and the patient.

Question no. 6: postoperative course has been simple, without facial palsy, but the patient has lost hearing in her left ear. What would you advise, given her job and her love of music? What results would you expect from fitting a hearing aid?

A. Chays

I have just one thing to advise: a hearing aid.

Several systems could be tried, without rushing but without waiting:

- a CROS system, with wifi or eyeglass routing;
- a vibrator on a diadem.

I would prepare the patient for these trials by explaining the why and the how. I would especially ask for her adherence in the various conditions of her sound environment. I would suggest a second consultation after the trials, in 2 or 3 months' time.

The upshot, from what I have seen with other patients, is unpredictable: some find such systems useless, while others are delighted, for no obviously apparent reason.

C. Dubreuil

She had lost her left ear before the operation.

Work-wise, I'm not sure she can still fly. But she could take a ground-staff post.

Listening to music at home, with good acoustics and good speakers, is often not a problem. At the opera or concert hall, on the other hand, it can be more complicated.

If the contralateral ear is good, that is often enough for listening to good music, especially as that ear shows no distortion.

A hearing aid — and it would have to be the CROS system — only serves to simulate stereophony, which is useful in society and certain work situations. This is not what the patient seems to be asking for just now.

I would wait and see before suggesting a hearing aid, which would lose a lot of the harmonics and interest of music: only work needs could determine the choice of fitting a hearing aid.

F.-M. Vaneecloo

A bone-implanted hearing aid, or a wifi CROS, could be suggested. The patient would need to be very carefully informed as to the advantages of the two methods: recovery of pseudo-stereophony. Stereo-audiometric tests with a vibrator on a headband are essential, to demonstrate the result to the patient. She needs to be able to think about the indication.

Currently, we suggest that this kind of patient undertake postoperative binaural listening rehabilitation.

J. Magnan

Flight staff count as security staff, and cophosis is an exclusion criterion. The occupational physician will need to reclassify her. Improved comfort of hearing requires rehabilitation of pseudo-stereophony. A bone-anchored hearing aid (BAHA), with a prior CROS trial to demonstrate the expected hearing gain, is indicated. The BAHA implant, however, involves a problem of esthetics, which is a limiting factor for many people.

Discussion

This third episode of so-called ''sudden'' deafness rightly worried the four experts, occurring as it did in a young woman who is both an air hostess and a musician, thus doubly concerned by her hearing.

One of the experts rightly suggested that a fuller assessment should have been conducted earlier. At all events, in case of ascending-type perceptual hearing loss, it is classical to look for triggering factors and to assess the role of context, and of stress in particular (even though the mechanism whereby stress affects hearing remains very unclear). Clinically, it is important to look for slight neurologic or vestibular signs, confirmed on complementary examinations performed the same day. Likewise, biological assessment of lipids, inflammation, hormones and hemostasis should be systematic. BAEPs, OAEs and ECOG are doubtless interesting, but in my own experience rarely shed real light.

The diagnosis suggested by the various experts was basically recurrence of cochlear hydrops this was supported by: the ascending auditory curve, highly suggestive of a pressure-related mechanism, the episodes of recurrence with apparently complete recovery, and the absence of any vestibular signs. A thrombotic accident of hormonal origin and intracochlear neurinoma were also suggested by one expert.

All, however, requested contrast-enhanced MRI, confirming the attitude that this examination should be systematic, although not urgent, in any case of sudden deafness, especially when recurrent. They were aware that an acoustic neurinoma-type tumoral process must always be investigated and ruled out, whatever the type of hearing loss.

The MRI performed following the episode did indeed reveal an intra-IAC neurinoma, involving the cochlear nerve according to some of them. The radiological features left no doubt as to the diagnosis. It was, however, crucial that at the far end of the IAC there was a layer of liquid significantly denser than the CSF of the pontocerebellar cistern: this doubtless accounted for certain aspects of the clinical history and, in particular, for the ascending pattern of the audiometric curve.

This density is due to an abnormally high protein concentration. This was first suspected more than 40 years ago by Georges Shambaugh, based on the yellowish color of the perilymph found on a translabyrinthine approach, and in the 1970s led certain authors to recommend perilymph sampling through the footplate as a diagnostic test for neurinoma. The supposed mechanism is that the tumor effectively plugs the IAC, preventing both normal circulation of the CSF and also, possibly, perilymph renewal. The intracochlear signal is often modified on T2-weighted sequences. Such abnormal density could account for pressure-related or metabolic hearing loss.

The experts also, however, raised another hypothesis: transitory ischemia or vasospasm of the labyrinthine and/or cochlear artery by labyrinthine artery compression in the IAC. Experimental work cited by one of the experts showed that temporary interruption of inner-ear arterial flow induced reversible suppression of cochlear nerve action potentials. It may be that mobilization and displacement of the neurinoma within the IAC can temporarily compress the labyrinthine artery, inducing transitory hearing loss.

Intralabyrinthine neurinoma extension (generally clear on MRI), iterative intratumoral hemorrhagic or inflammatory phenomena or iterative work-related dysbaric events, on the other hand, appeared less relevant.

At all events, these episodes of sudden deafness were of poor prognosis for surgical conservation of hearing function.

Therapeutically, treatment options seemed straightforward, inasmuch as the patient declined MRI surveillance and wanted to get rid of the tumor ''in her brain''.

Surgical exeresis with a translabyrinthine or retrosigmoid approach was envisaged, either to try to conserve hearing for the first expert or to sacrifice it deliberately, given the poor chances of conservation, for the third. Both, however, insisted on the necessity of informing the patient as to the risk, not so much of facial palsy, although this can never be zero, as of loss of hearing.

At all events, all four experts rejected stereotactic radiotherapy: long-term doubts as to the evolution of this very small target tumor and the risk of long-term hearing impairment were the central arguments.

Even so, two of the experts stressed the interest of simple radiologic surveillance for a woman who is both young and a musician. The arguments are far from negligible: a risk of jeopardizing both her job and her center of interest in case of postoperative cophosis, the rather minor inconvenience represented by moderate left hearing loss, and the feasibility of precise MRI monitoring of tumor evolution. As one of them puts it, ''the treatment should not be more aggressive than the tumor image''. Exhaustive discussion to explore the patient's motives and provide ''enlightened'' information was thus essential before surgery can be suggested.

And, in fact, this discussion would seem to have proved fruitful, inasmuch as the patient finally opted for simple surveillance. One year on, however, the question no longer arose. Surgery had become fully justified, as threat to the hearing function no longer applied, the patient having become ''spontaneously'' cophotic.

One expert recommended preoperative electrophysiological assessment of the facial nerve, given the atypical nature of the clinical history. In point of fact, such peroperative assessment is classical (and may yet become mandatory). It may reveal infraclinical signs of facial involvement, in which case the patient should be informed as to the increased risk of postoperative facial palsy. In my own as in the other experts' experience, such assessment does not seem obligatory in case of intracanal tumor, partly because genuine facial nerve schwannoma is associated with normal electrophysiological findings, and also because abnormal electrophysiological responses are not systematically associated with facial nerve involvement.

The recommended approach seemed to be preferentially translabyrinthine, due to the preoperative cophosis, the basically otological technique and ease of control of the facial nerve in the IAC.

In the present case, surgery was in fact performed without difficulty and postoperative course was simple.

Two issues remain: possible rehabilitation of hearing, and the patient's future work.

Three experts recommend a CROS or BAHA hearing aid, while stressing the need for prior trials and assessment and also the limitations and drawbacks. Results indeed would seem to be unforeseeable from one patient to another, and at all events the functional improvement obtained would at best consist in pseudo-stereophony, which is of limited interest for a musician. The BAHA also raises practical and esthetic problems, which the last expert was right to underline.

For my part, I would share the attitude of the second expert: abstention. Apart from the issues raised above, there is also that of the ''visibility'' of auditory disability in this woman who is an air hostess, and hence of her future career.

The experts envisage reclassification to ground staff; but there is in fact a certain fuzziness in the regulations in force.

In practice, flight staff, including stewards, must undergo a two-yearly medical check-up. This is performed in approved expert centers and, in theory, includes audiometry; should the audiogram show significant evolution since the previous examination, the file is sent on to the civil aviation medical council (*Conseil médical de l'aviation civile* [CMAC]), who undertakes a fresh assessment on the basis of which an opinion is formulated: renewed or restricted flight authorization, temporary suspension, restriction to ground staff, etc.

Decision criteria, however, appear to be various: degree of hearing loss; real or subjective difficulty in work; disturbed balance; associated tinnitus or facial palsy; patient complaints, motivation and attitude to reclassification, etc. In other words, and however surprising it may seem, cophosis does not in itself contraindicate an aptitude to fly.

Conflict of interest statement

No conflict of interest.