CASE REPORT

Rare cause of bilateral sudden deafness

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SUMMARY

In this paper, we describe the case of a 62-year-old female with recurring episodes of sudden deafness with vertigo and facial paresis. Within a month's time, this resulted in bilateral deafness and vestibular areflexia. Erroneously, the patient was diagnosed with sudden deafness of unknown origin and subsequently with neuroborreliosis (Lyme disease). The true diagnosis of relapsing polychondritis (RP) was revealed 9 months after initial presentation. The diagnostic delay is in part explained by the fact that, by definition, the disease has to relapse before the diagnosis can be made, but also by its pluriform clinical presentation. Timely identification of RP as the cause of this profound sensorineural hearing loss proved to be important. It was key in instigating adequate follow-up, and allowed for cochlear implantation before total cochlear obliteration, which might have hampered optimal hearing rehabilitation.

BACKGROUND

Sudden deafness is a term used for unexplained, rapid (mostly sensorineural) hearing loss of >30 dB, which occurs over less than a 72-hour period.¹ It usually affects one ear, and simultaneous or subsequent involvement of the contralateral ear is rare. Even though a plethora of possible causes like infectious labyrinthitis, microvascular events and autoimmune disorders is proposed, aetiology remains unresolved in many cases. Standard empirical treatment of idiopathic sudden deafness includes prednisolone, either orally or intravenously.² Some episodes of sudden deafness, however, result in permanent, profound, hearing loss despite this therapy.

The correct identification of the cause of sudden hearing loss is important, even though the audio-vestibular damage may be irreversible. It facilitates adequate follow-up and optimal hearing rehabilitation and may aid in a correct prognosis concerning the ipsilateral as well as the contralateral side. This case illustrates the clinical relevancy and difficulties in making the right diagnosis.

CASE PRESENTATION

A 62-year-old female with hypertension and a single episode of uveitis in her medical history presented to the department of otolaryngology of a neighbouring hospital with the symptom of sudden deafness, tinnitus on the right side and vertigo. Otoscopy showed no abnormalities and facial nerve function was intact. MRI (figure 1) showed contrast enhancement of the labyrinth (the vestibule and cochlea), a sign of inflammation. The patient was diagnosed with sensorineural hearing loss caused by autoimmune inflammation or infectious labyrinthitis and was treated with prednisolone orally.

Figure 1 T1 contrast-enhanced MRI performed after initial presentation with sudden deafness, tinnitus and vertigo on the right side. Enhancement of the right labyrinth (cochlea and vestibule) can be observed as a sign of autoimmune or infectious inflammation.

The symptoms of tinnitus and vertigo improved gradually while the deafness persisted. A month later, the patient presented with sensorineural hearing loss on the (contralateral) left side, accompanied by left-sided facial paresis (House Brackmann grade IV). Further physical examination was normal. A second MRI scan (figure 2) showed contrast enhancement of the left labyrinth, similar to what had previously been observed on the right side. The patient had not experienced a recent episode of erythema migrans; however, the possibility of Lyme borreliosis (LB) was considered because of contralateral involvement of the labyrinth and simultaneous facial nerve paresis. Serum analysis for LB was negative for IgG, but positive for IgM antibodies. Spinal cord fluid showed no direct signs of LB, but a leucocytosis (32 cells/ μ L) and elevated proteins (710 mg/L) were observed.

Figure 2 Second T1 contrast-enhanced MRI, 1 month after initial presentation, performed during second episode of sudden deafness on the contralateral (left) side. Again, enhancement of the labyrinth (cochlea and vestibule) can be observed as a sign of autoimmune or infectious inflammation.

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The patient was additionally evaluated by the neurologist, and the diagnosis was altered to bilateral sudden deafness and facial paresis most probably caused by (early) LB. The patient was subsequently treated for neuroborreliosis with intravenous antibiotics (ceftriaxone) during 14 days. Post-treatment hearing tests revealed severe bilateral sensorineural hearing loss and electrocochleography showed bilateral vestibular areflexia. Although facial nerve function normalised in the months after this episode, the deafness remained and the patient was referred to our Department of Otolaryngology-Head and Neck Surgery. The hearing loss was so severe that the criteria for cochlear implantation were met, and this procedure was subsequently performed on the left side. During the procedure, the base of the cochlea was found to be partly calcified, most probably due to postinflammatory changes of the cochlear lumen. In this case, however, the cochlear implant could be fully inserted and the procedure was otherwise uncomplicated. However, 4 days after cochlear implantation, the patient presented with redness and swelling of the pinna of the operated left ear without involvement of the incision line or earlobe, indicative of chondritis. In retrospect, the patient remembered that the first episode of sudden deafness on the right side had been accompanied by similar redness and swelling of the right ear (figure 3). No diagnosis causing the redness was made during this past episode, as the perichondritis soon resolved spontaneously and she considered the episode to be of no significance. It was therefore not included in the list of symptoms leading to the previous differential diagnosis of idiopathic sensorineural hearing loss and neuroborreliosis. Although the presentation during the current episode was not typical for a wound infection and appeared to be chondritis, this possibility could not be ruled out and the

patient was treated with amoxicillin and clavulanic acid orally. The redness and swelling subsided within 2 weeks after presentation. Repeat serum analysis for LB showed no seroconversion, that is, IgG antibodies remained negative.

DIFFERENTIAL DIAGNOSIS

Although sudden deafness, facial paresis, vertigo and tinnitus can all be attributed to LB, chondritis of the pinna cannot, and the lack of LB seroconversion made this diagnosis unlikely. Also, enhancement of the cochlea on MRI and the intraoperatively observed calcification of the cochlea are signs of an infection of the labyrinth, not of the vestibulocochlear nerve. This makes the diagnosis of LB less likely, as (neuro) LB is most commonly known to affect cranial nerves. Review of all symptoms that had occurred over time, including two episodes of chondritis and an episode of uveitis in the patients' history, prompted the diagnosis of relapsing polychondritis (RP). To confirm this diagnosis, ANA and ANCA antibodies were evaluated and a biopsy of the left pinna was taken. However, ANA and ANCA antibodies were not elevated and the histology showed no signs of chondritis. Even so, all clinical criteria necessary to officially diagnose RP were met and the patient was diagnosed having RP.

TREATMENT

As the inflammation of the ear had subsided, no further treatment of RP was deemed necessary.

OUTCOME AND FOLLOW-UP

The patient has been free of symptoms for the past 14 months, and hearing rehabilitation has been satisfactory with the cochlear implant.

DISCUSSION

This case illustrates that affirming RP as the cause of sudden deafness can be challenging. Especially when it is accompanied by rare symptoms like facial paresis, a faulty initial diagnosis like LB is easily made. One of the main reasons why LB was considered in this case was because of the positive laboratory result for borreliosis IgM antibodies. However, the use of IgM antibodies has a high false-positive rate, as 5–10% of all people will test positive without actually having the disease.⁵ Furthermore, concurrent autoimmune disease, like RP, is known to even further increase false-positive test results for borreliosis IgM antibodies.³

RP is a rare and potentially fatal autoimmune disorder that affects cartilaginous structures and connective tissue and has a pluriform clinical presentation. It affects ~1 in 3.5 million people per year.⁴ The time of diagnosis is mostly in the fifth decade of life, but the first signs of RP can present from childhood to old age with a female predominance.⁴ Auricular chondritis, arthritis and respiratory tract involvement are the most common signs in RP,⁵ followed by ocular inflammation and nasal chondritis.⁶ Mucocutaneous, haematological, cardiovascular, neurological and renal manifestations are also reported in patients with RP. Sensorineural hearing loss and vestibular dysfunction, as observed in this case, are less common, and facial nerve involvement is rare.^{6,7} It is important to recognise RP as a possible cause of sudden deafness. Postinflammatory calcification of the cochlea after an episode of RP, like we found in our patient, has rarely been described, but may hamper cochlear implantation and optimal hearing rehabilitation. Furthermore, in case of RP, it is necessary to evaluate other organ systems that might be affected and to provide adequate follow-up.

Figure 3 The right ear of the patient at the time of initial presentation. The ear is red and swollen and accompanied by symptoms of sudden deafness, tinnitus and vertigo.

RP is a clinical diagnosis, based on a set of clinical criteria (first described by McAdam *et al*⁸ and later modified by Damiani and Levine,⁹ box 1).

There are no serological or histological tests that can unequivocally prove the diagnosis. Furthermore, it is not possible to make a definite diagnosis during the first symptomatic episode of the disease, as episodes must 'relapse' in order to qualify as RP. This can result in a delay in diagnosis, as was the case in our patient, with a reported median delay of 3–5 years⁴ after the first reported symptoms. Circulating autoantibodies like ANA and ANCA have a limited added diagnostic value, as positive test results can often be related to other associated autoimmune diseases, and negative test results (like in our patient) are observed in over 50% of patients with RP.⁶ Histological confirmation of chondritis in a biopsy sample can contribute to the diagnosis of RP and is one of the non-clinical criteria for RP as proposed by Damiani and Levine.⁹ In the current case, chondritis could not be histologically confirmed, possibly due to the small size of the biopsy (2 mm of cartilage) or the timing of the biopsy (10 days after the first symptom of a swollen and red ear). All the same, the clinical criteria for RP were met convincingly when reviewing all clinical features that had occurred over time.

There is no definitive cure for RP. The prognosis of RP depends on the site of inflammation. The management of RP focuses on relieving symptoms and prevention of complications. Peripheral arthritis and (auricular or nasal) chondritis can be treated with low-dose corticosteroids. Treatment with high-dose corticosteroids is used in cases of severe disease manifestations and surgery might be required when there is cardiac involvement.⁶ Inflammatory involvement of the respiratory tract, cardiac valvular disease and advanced systemic vasculitis have been reported as major causes of mortality. As the management of complications has improved over the past decades, so has the 10-year survival rate.⁵

Bilateral sensorineural hearing loss in RP has been described before; however, this case is one of very few in which a cochlear implant was indicated after sudden deafness caused by RP.^{10–12} Cochlear implantation should be considered when profound bilateral sensorineural hearing loss has occurred that does not respond to conservative therapy. Profound and stable hearing

loss should, in our opinion, be the final indication to perform cochlear implantation, independent of radiological findings on subsequent CT or MRI. The resolution of the intracochlear inflammatory process does not result in increased hearing and does not predict whether or not the cochlea will become obliterated by ossification or not. In fact, cochlear ossification as a rule is preceded by a resolution of the contrast enhancement that is indicative of cochlear inflammation on MRI.¹³ However, when signs of postinflammatory calcification of the cochlea are identified on CT or MRI, referral to a cochlear implant centre as soon as possible is recommended in order to enable cochlear implantation before further calcification of the cochlea has occurred. If not, postinflammatory calcification of the cochlea may prevent successful cochlear implantation, thereby precluding optimal hearing rehabilitation.¹⁴ Our recommendation therefore is to urgently refer any patient with bilateral sudden deafness to a cochlear implant centre, especially when an inflammatory process like RP is thought to be the causative pathology.

Owing to the pluriform presentation and relapsing nature of RP, patients almost never present with the 'full clinical picture' of RP. Because of this, different doctors of different disciplines (mostly general practitioners, otolaryngologists, ophthalmologists and rheumatologists) see different symptoms at different moments in time. Frequently, symptoms have initially been attributed to other forms of disease, and only careful history taking with attention to symptoms beyond the scope of one's own specialty will reveal the diagnosis.

Learning points

- ▶ Bilateral sudden deafness is rare and should prompt thorough clinical investigation.
- ▶ Relapsing polychondritis (RP) is a possible cause of sudden deafness. The diagnosis is based on clinical signs and symptoms as there is no serological or histological test that can definitely prove the diagnosis.
- ▶ The most common features of RP are auricular and nasal chondritis, audiovestibular dysfunction and ocular inflammation.
- ▶ Owing to its pluriform presentation over time, it is very likely that different doctors of different disciplines have already reviewed and diagnosed the patient, before the definitive diagnosis of RP is made.
- ▶ If profound sensorineural hearing loss occurs as a result of RP, it is essential to swiftly perform adequate imaging of the cochlea, as postinflammatory calcification of the cochlea might hamper cochlear implantation and thus preclude optimal hearing revalidation.

Box 1 Diagnostic criteria for relapsing polychondritis. Established by McAdam *et al*⁸ and later adjusted by Damiani and Levine⁹

Clinical features:

- ▶ Bilateral auricular chondritis.
- ▶ Non-erosive, seronegative inflammatory polyarthritis.
- ▶ Nasal chondritis.
- ▶ Ocular inflammation (conjunctivitis, keratitis (epi)scleritis, uveitis).
- ▶ Respiratory tract chondritis.
- ▶ Cochlear or vestibular dysfunction or both.

Diagnosis (one of the following):

- ▶ Three clinical features.
- ▶ One clinical feature and histological confirmation of chondritis.
- ▶ Chondritis at two or more anatomical locations, with positive response corticosteroids, dapsons or both.

Competing interests None declared.

Patient consent Obtained.

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