Effect of rituximab on AAV-related otitis media case

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Introduction

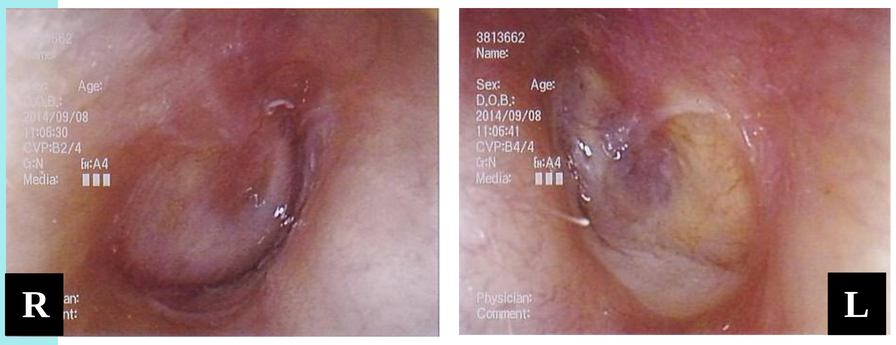
The combination of glucocorticoids and cyclophosphamide leads to remission in most patients with anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV)-related otitis media. However, complete deafness cannot be reversed, and the relapse rate remains high even when maintenance treatment with immunosuppressive drugs is administered. We review the diagnostic features of AAV-related otitis media and discuss the effect of rituximab (a molecularly targeted drug) as a substitute for cyclophosphamide.

Case report

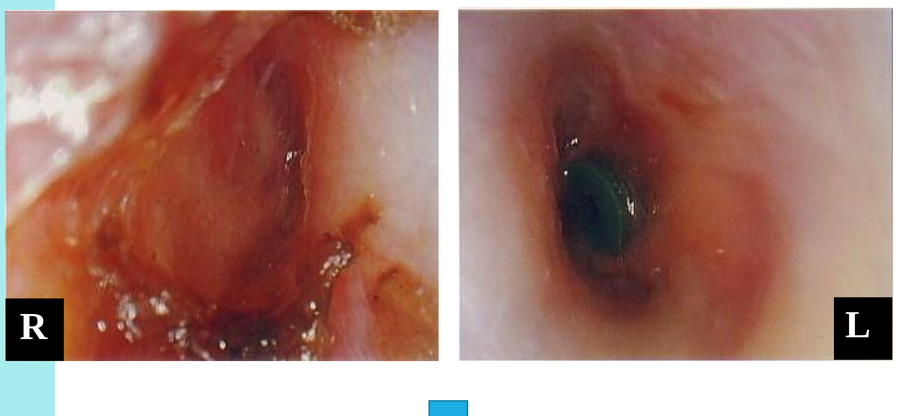
We reviewed the case of myeloperoxidase (MPO)-ANCA-positive patient who initially complained of otologic symptoms and was referred to Fukushima Medical University.
Patient: An 81-year-old woman was referred to us with a high MPO-ANCA titer and bilateral hearing loss with middle ear effusions, after 6 months of treatment for otalgia. She had almost complete deafness.
The following findings were observed: positive MPO-ANCA (279 EU/ml [normal <3.5 EU]), negative proteinase 3 (PR3)-ANCA (<10 EU), and bilateral hearing loss (average hearing level: right, 105 dB; left, 101 dB). Following a tapered course of prednisolone (initial dose 40 mg/d) and subsequent initiation of rituximab at 557 mg/d, her MPO-ANCA titer returned to 1.7 EU/ml and her bilateral hearing loss showed marked improvement (right, 56.3 dB; left, 57.5 dB). No relapse occurred.

Findings of the eardrum

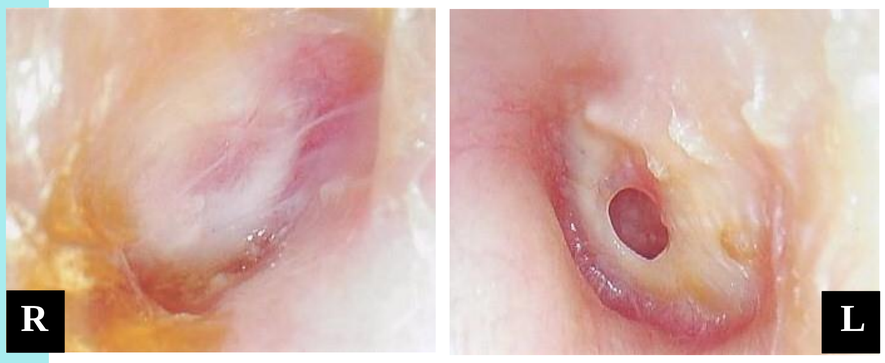
(A) First examination



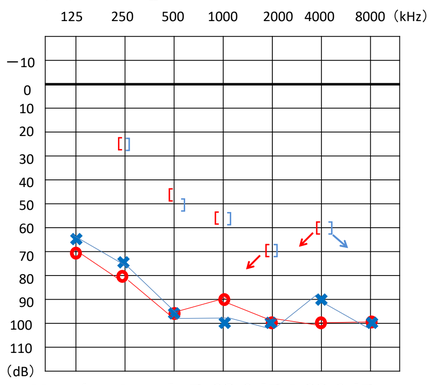
(B) Otalgia & Fever onset



(C) After treatment

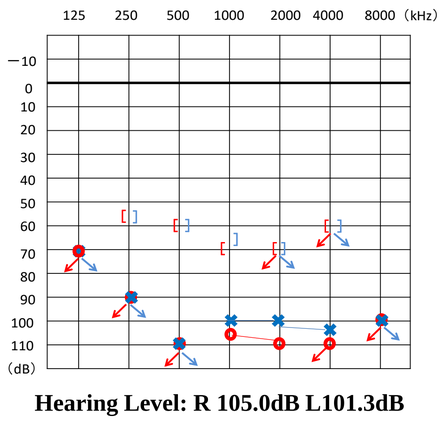


(A)audiogram before treatment



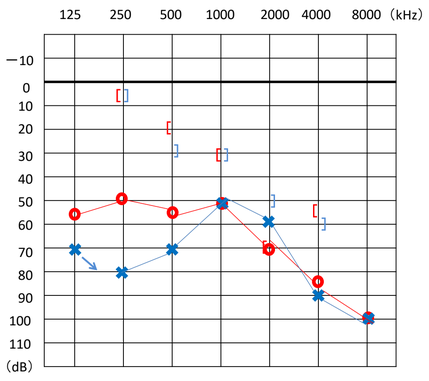
Hearing Level: R 93.0dB L92.0dB

(B)audiogram before treatment



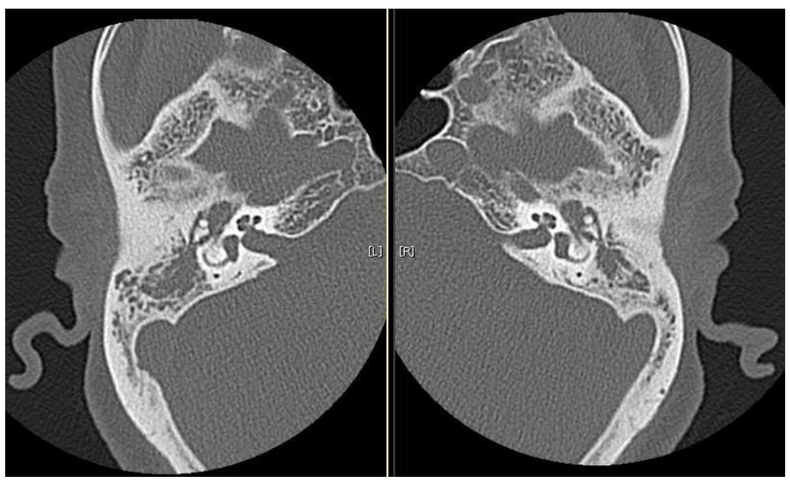
Hearing Level: R 105.0dB L101.3dB

(C)audiogram after treatment



Hearing Level: R 56.3dB L57.5dB

temporal bone computed tomography



laboratory findings

WBC 8800/μl

CRP 1.87 mg/dl

T-SPOT.TB (−)

PR3-ANCA (−)

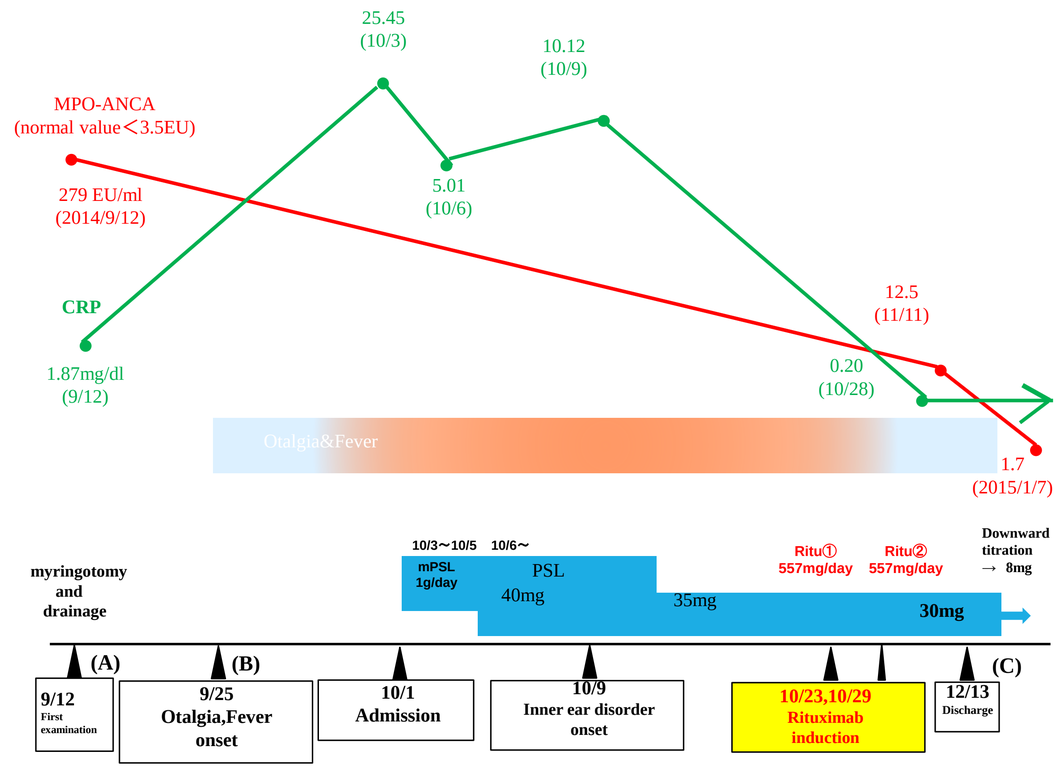
MPO-ANCA 279 EU/ml

(normal value<3.5 EU/ml)

＊Facial palsy(−)

＊No upper respiratory tract lesion except the ear

clinical course after first examination



Discussion

The combination of cyclophosphamide and glucocorticoids leads to remission in most patients with ANCA-associated vasculitides. However, even when patients receive maintenance treatment with immunosuppressive drugs, the relapse rate remains high. Using Rituximab in substitution for an immunosuppressive drug may help to maintain remission[1)] .

Rituximab, a chimeric anti-CD20 monoclonal antibody, has been shown to be quite effective in the treatment of immune disorders resulting from autoantibodies[2)]. It is also an effective and well-tolerated treatment for patients with AAV and should be strongly considered in severely affected patients who do not respond to standard therapy or in those in whom cytotoxic therapy bears a high risk morbidity.

Yoshida et al. reported that after starting immunosuppressive therapy, ANCA titers rapidly returned to the normal range and hearing levels improved, but the complete deafness could not be reversed[3)]. In the present case, hearing level recovered from almost complete deafness after administration of rituximab.

Yoshida et al. also reported that the early ‘reversible’ state of sensorineural hearing loss might be caused by a reduction in K^+ ions in the endolymph due to dysfunction of the stria vascularis where the microcirculation networks are well developed in the cochlea, this sustained microvasculitis might lead to dysfunction of fibrovascular coupling following permanent ‘irreversible’ damage of hair cells[4)]. In this case, the function of stria vascularis might be kept well, so hearing level recovered from almost complete deafness after administration of rituximab.

Rituximab maintenance therapy was introduced due to being old, a steroid-resistant thing, condition progressing, and the disorder being more likely to recur. As a result, the symptom is improved (in acknowledgment of particularly aural recovery), and there is not the relapse of the symptom now either. The possibility that Rituximab was useful for treatment of ANCA-related vasculitis-related otitis media was suggested.

Conclusions

This case study showed the effect of rituximab on AAV-related otitis media. Further cases should be prospectively examined and reviewed to establish the treatment for AAV-related otitis media.

References

1)Guillevin, et al. 2014 N Engl J Med. 2)Stasi, et al. 2006 Rheumatology. 3)Yoshida N, et al. 2014 Otology & Neurotology. 4)Yoshida N, et al. 2014 Allergology International.

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