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Case reports

A solely ear-involved IgG4-related sclerosing disease with two-years following-up

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ABSTRACT

Introduction: Immunoglobulin G4-related sclerosing disease (IgG4-RSD) is a chronic fibro-inflammatory disease involving systemic multi-organ lesions, such as the salivary and lacrimal glands, lymph nodes and pancreas. The diagnosis of this disease is reliant upon clinical manifestations, laboratory tests, histopathologic results and radiological data. Some studies have found that IgG4-RSD has otological manifestations, whereas there were few studies introducing the diagnosis, therapy and long-term follow-up results of solely otological IgG4-RSD.
Case summary: Here, we report a case of IgG4-RSD involving the ear alone. A female presenting with otalgia and hearing loss underwent surgery, without hormone therapy. The pathological diagnosis was otological IgG4-RSD and no clinical or radiological signs of recurrence were observed at seven and twenty-four months follow-up.
Discussion: This case indicates that IgG4-RSD can invade the ear only, and that surgical therapy without hormone therapy is efficient for solely IgG4-RSD. Pathological results are crucial for diagnosis.

1. Introduction

Immunoglobulin G4-related sclerosing disease (IgG4-RSD) is a systemic entity that usually causes irreparable damage to the pancreas, salivary glands, kidney, gastrointestinal tract, eyes, lymph nodes, and other organs [1]. IgG4-RSD is hard to diagnose because the clinical symptoms cannot be found until organs undergo swelling or damage. By far, some studies have shown otological involvement of IgG4-RSD, the clinical manifestations of which are mainly hearing loss and otitis media [2,3]. Few cases of solely otological involvement have been previously reported. Here, we report a case of IgG4-RSD with exclusively otological involvement that was treated surgically and followed up for two years.

2. Case report

A 54-year-old female patient presented with a 10-year history of otalgia, tinnitus and progressive hearing loss in the left ear. She had also experienced a few episodes of whirling-type dizziness. An otoscopic examination showed an intact left tympanic membrane with an obvious bulge in the pars flaccida (Fig. 1A). Pure tone audiometry was normal in the right ear, whereas the hearing level of the left ear decreased and the pure tone average at 500 Hz to 2 kHz frequencies was 86.7 dBHL (Fig. 1B). Computed tomography (CT) of the middle ear revealed chronic left middle ear mastoiditis with accompanying cholesteatoma in the tympanic cavity and antrum, as well as partial bone erosion (Fig. 2A). Magnetic resonance imaging (MRI) of the middle ear showed an enhanced area of abnormal soft tissue occupying the left tympanic cavity and antrum (Fig. 2B,C). The MRI results suggested a granulomatous inflammatory or tumorous lesion in the middle ear.

A left mastoidectomy was performed for biopsy and lesion tissue was totally removed. The pathological results of the submitted tissues revealed dense lymphoplasmacytic infiltrate (Fig. 3A), storiform-type fibrosis (Fig. 3B) and obliterative phlebitis (Fig. 3C) in the tumor. Immunohistochemical findings showed: vimentin(+), lysozyme(part +), CD68(part +), Cdla(−), S100(+), Ki67(5% +), CD34(vessel +), EMA(+), Ckpan(−), Syn(−), NSE(−), IgG(part +) and IgG4(+). The ratio of IgG4-positive plasma cells to IgG-positive plasma cells was > 40% and IgG4-positive plasma cells per high-power field > 10 (Fig. 3D). Thus, the pathological diagnosis was IgG4-RSD.

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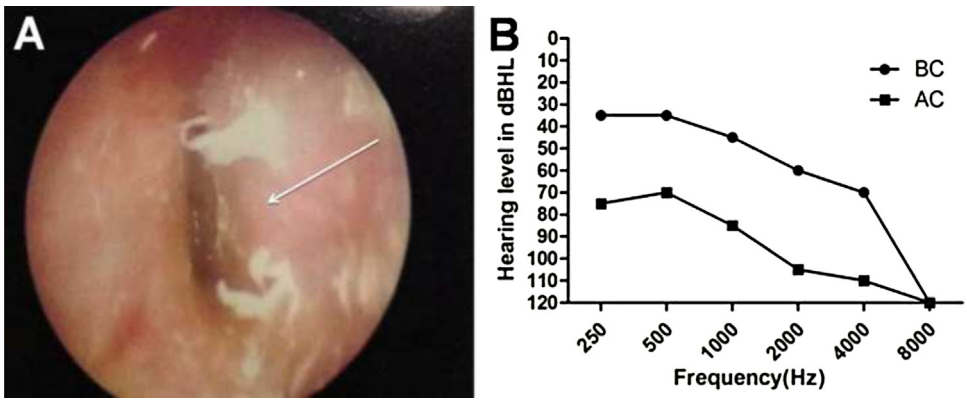

Fig. 1. A: Otoscopy of the left ear of our patient. An obvious bulge is seen in the pars flaccida. The white arrow indicates the lesion site; B: Pure tone audiogram of the left ear.

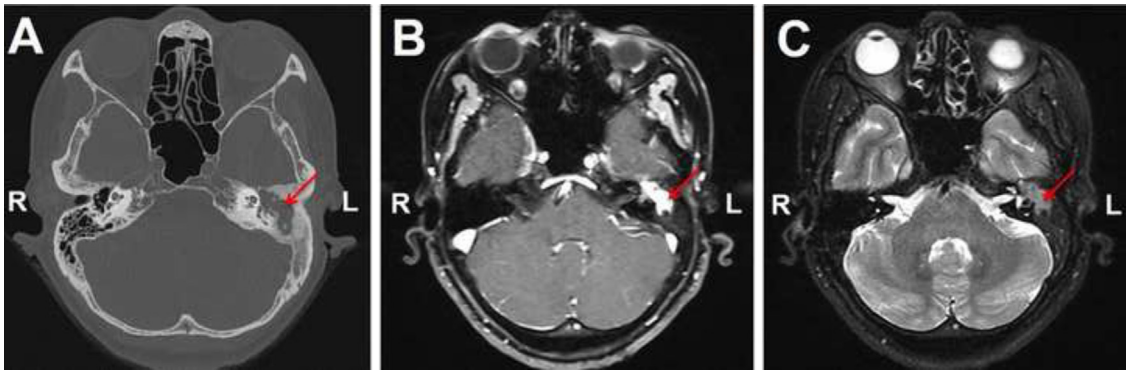

Fig. 2. A: CT with bone windows; B: contrasted T1-weighted MRI; C: T2-weighted MRI of the temporal bone preoperatively. The red arrows indicate the lesion site.

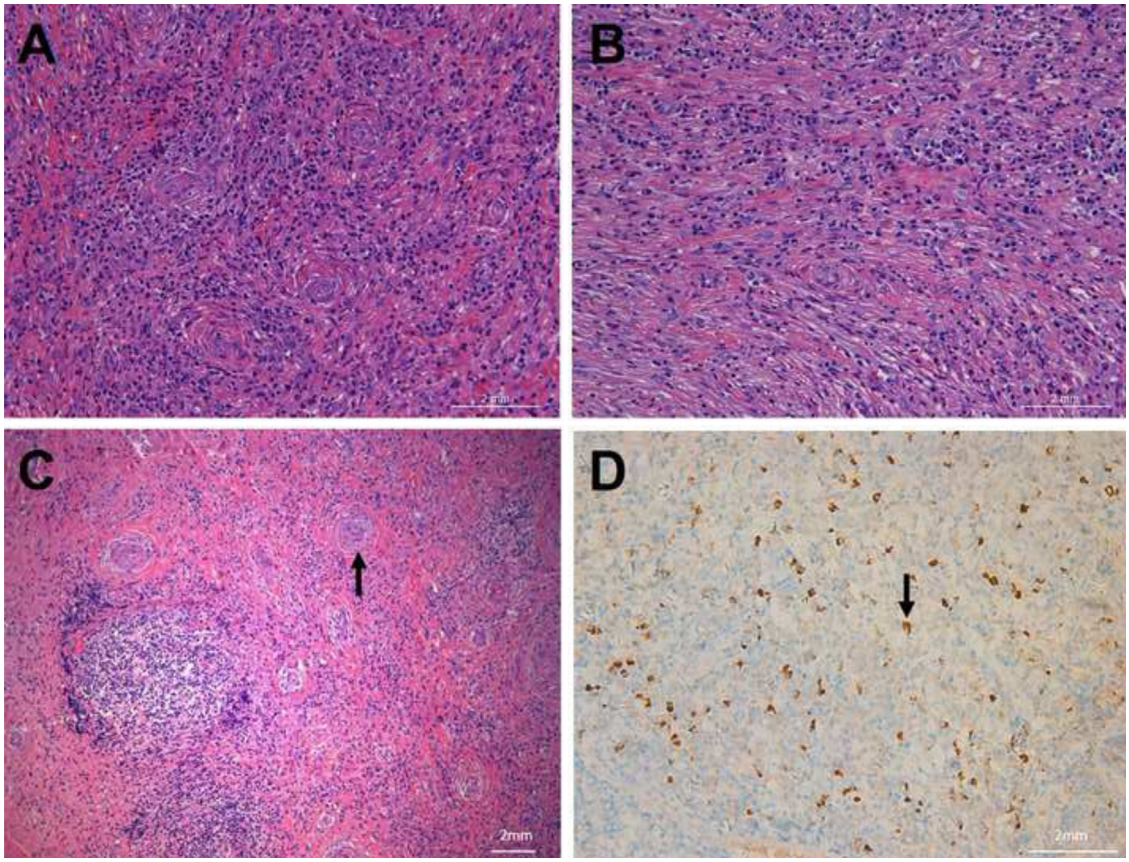

Fig. 3. Hematoxylin and eosin and immunological staining of the tympanic cavity tumor in the left ear. A: A dense lymphoplasmacytic infiltrate (original magnification × 20); B: Storiform-type fibrosis in the tumor (original magnification × 20); C: Obliterative phlebitis in the tumor (black arrows; original magnification × 10); D: Multiple IgG4-positive plasma cells (black arrows; original magnification × 20) demonstrated by immunostaining for IgG4.

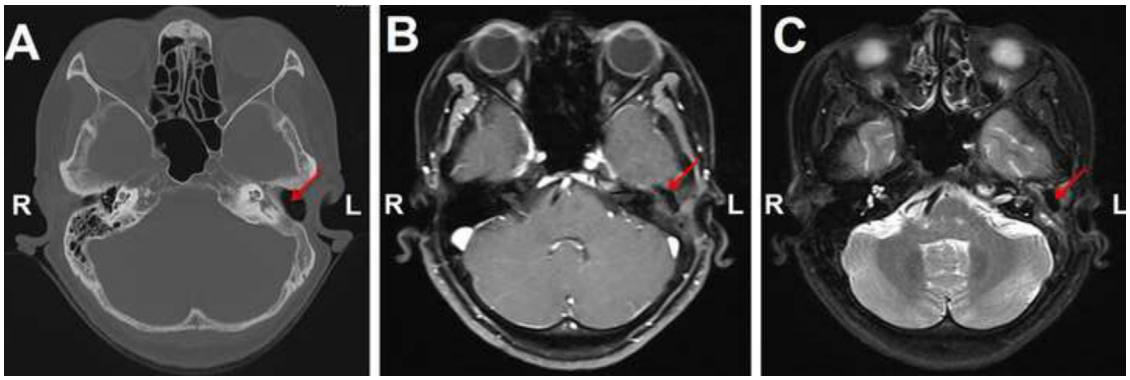

Fig. 4. CT with bone windows, contrasted T1-weighted MRI and T2-weighted MRI of the temporal bone obtained at A: twenty-four; B,C: and seven months postoperatively. The red arrows indicate the lesion site.

The patient underwent a complete imaging workup ination which included neck and abdominal B-ultrasonography and head and chest CT. The results showed no other organic manifestations except for middle ear. One week after operation, the serum IgG4 concentration of this patient was examined and the IgG4 concentration was not elevated. Therefore, the patient did not receive hormone therapy. Serum IgG4 concentration remained normal at seven and twenty-four months after operation. The reexamination of CT and MRI (Fig. 4A–C) showed that the lesion was removed compared with preoperative imaging results. The patient recovered smoothly and no clinical and radiological signs of recurrence were observed at seven and twenty-four months follow-up.

3. Discussion

IgG4-RSD is an immune-mediated disease characterized by tumorous swelling of damaged organs and an elevated serum IgG4 concentration [4]. The pancreas, kidneys, lymph nodes, parotid glands, eyes, and retroperitoneum are the organs primarily affected by IgG4-RSD and are often irreversibly damaged [5]. There are three key findings of histopathological features relevant to the diagnosis of IgG4-RSD: dense lymphoplasmacytic infiltrate, storiform-type fibrosis and obliterative phlebitis [5,6]. This case of histopathological results also showed that the ratio of IgG4-positive plasma cells to IgG-positive plasma cells was > 40% and IgG4-positive plasma cells per high-power field > 10. The histopathological results met the diagnostic criteria for IgG4-RSD. In this case, the results of CT and MRI showed that there were masses located in the middle ear cavity which also supported the diagnostic criteria.

Otological involvement of IgG4-RSD has been reported in previous studies [2,3,7,8], but the involvement was usually accompanied by other organic manifestations. In this patient, the ear was the only organ invaded by IgG4-RSD and the results of full examination showed no evidence of extra-ear organ involvement. CT and MRI revealed localized masses accompanied by bone erosion which resembled cholesteatoma, granulomatous inflammation or a tumor lesion. The serum IgG4 concentration of patients with IgG4-RSD is elevated, but varies widely. Elevation of the serum IgG4 concentration aids the diagnosis of IgG4-RSD [9], and can be used to further validate the diagnostic results, monitor the therapeutic process, and evaluate the therapeutic effect [10]. The preoperative serum IgG4 and IgE concentrations of this case were not examined, but at one week, seven and twenty-four months after surgery, the concentrations were normal. The results suggested that IgG4-RSD involved the ear only, or affected the ear first and does not influence the serum IgG4 concentration. Otherwise, it also may be that the concentration of serum IgG4 decreased at one week after surgery.

This patient had received surgical therapy to entirely remove the mass and did not receive hormone therapy after the surgery. At seven and twenty-four months after surgery, there were no signs of relapse upon reexamination. These results suggest that surgical operation without hormone therapy is efficient for treating IgG4-RSD that involves the ear exclusively.

4. Conclusion

In conclusion, IgG4-RSD can invade the ear exclusively, which is associated with early and clinical manifestations of long-term otalgia and progressive hearing loss. Histopathological examination remains the gold criterion for a confirmed diagnosis. Surgery without hormone therapy was efficient for solely otological IgG4-RSD.

Disclosure of interest

The authors declare that they have no competing interest.

References

[1] Khosroshahi A, Wallace ZS, Crowe JL, Akamizu T, Azumi A, Carruthers MN, et al. International Consensus Guidance Statement on the Management and Treatment of IgG4-Related Disease. Arthritis Rheumatol 2015;67:1688–99.
[2] Bittencourt AG, Pereira LV, Cabral Jr F, Halang Fde S, Goncalves Mde C, Bento RF. IgG4-related sclerosing disease of the temporal bone. Otol Neurotol 2013;34:e20–1.
[3] Takagi D, Nakamaru Y, Fukuda S. Otologic manifestations of immunoglobulin G4-related disease. Ann Otol Rhinol Laryngol 2014;123:420–4.
[4] Hamano H, Kawa S, Horiuchi A, Unno H, Furuya N, Akamatsu T, et al. High serum IgG4 concentrations in patients with sclerosing pancreatitis. N Engl J Med 2001;344:732–8.
[5] Kamisawa T, Zen Y, Pillai S, Stone JH. IgG4-related disease. Lancet 2015;385:1460–71.
[6] Deshpande V, Zen Y, Chan JK, Yi EE, Sato Y, Yoshino T, et al. Consensus statement on the pathology of IgG4-related disease. Modern pathology: an official journal of the United States and Canadian Academy of Pathology Inc, 25; 2012. p. 1181–92.
[7] Cho HK, Lee YJ, Chung JH, Koo JW. Otologic Manifestation in IgG4-Related Systemic Disease. Clin Exp Otorhinolaryngol 2011;4:52–4.
[8] Schiffenbauer AI, Wahl C, Pittaluga S, Jaffe ES, Hoffman R, Khosroshahi A, et al. IgG4-related disease presenting as recurrent mastoiditis. Laryngoscope 2012;122:681–4.
[9] Umehara H, Okazaki K, Masaki Y, Kawano M, Yamamoto M, Saeki T, et al. Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD), 2011. Mod Rheumatol 2012;22:21–30.
[10] Carruthers MN, Khosroshahi A, Augustin T, Deshpande V, Stone JH. The diagnostic utility of serum IgG4 concentrations in IgG4-related disease. Ann Rheum Dis 2015;74:14–8.