European Annals of Otorhinolaryngology, Head and Neck diseases 136 (2019) 199–201

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Case report

Hypereosinophilic syndrome presenting with bilateral ear fullness

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ARTICLE INFO

Keywords:
Hypereosinophilic syndrome
Otitis media
Eosinophilia

ABSTRACT

Introduction: Hypereosinophilic syndrome is a rare disease with hypereosinophilia resulting in end-organ dysfunction. Patients present with organ-associated symptoms, and the targets frequently affected are heart, lung, skin, or the nervous system, and the middle ear involvement is rare.
Case report: A 30-year-old female with left ear fullness and hearing loss, which persisted for 6 months, was finally diagnosed with hypereosinophilic syndrome (HES). After high dose systemic steroids treatment, all symptoms improved.
Conclusion: Eosinophilic otitis media and HES involving the middle ear share many clinical manifestations. Prompt and accurate differential diagnosis is required for these diseases to ameliorate symptoms and promote recovery.

1. Introduction

Hypereosinophilic syndrome (HES) is characterized by abnormal accumulations of eosinophils in peripheral tissue or the bloodstream. Idiopathic HES can be diagnosed when eosinophils accumulate to more than 1500 cells/μL in the blood without other disorders associated with hypereosinophilia [1], such as parasite infection, allergic reaction to drugs, malignancies, and autoimmune diseases. HES usually involves the cardiovascular system, respiratory system, skin tissue and nerves, and has only rarely been reported to involve the middle ear. In fact, only two such cases have been previously reported [2,3], and involvement of only the middle ear in HES has not been reported. Here we describe our experience of HES case involving only the middle ear.

2. Case presentation

A 30-year-old female with left ear fullness and hearing loss, which persisted for 6 months, visited our clinic. She had been diagnosed with left otitis media and had taken antibiotics for 2 weeks, but her symptoms did not improve.

At the initial visit, endoscopic examination revealed a bulging left tympanic membrane. Examination of the nasopharynx was unremarkable. Right pure tone threshold was 5 dB (average threshold at 500 Hz, 1000 Hz and 2000 Hz) and the left ear exhibited a 25 dB air-bone gap (bone conduction threshold 5 dB, air conduction threshold 30 dB) by pure tone audiometry (PTA). A viscous material was discharged with left myringotomy and ventilation tube insertion was performed on the left tympanic membrane under the diagnosis of middle ear effusion. She was not followed up for 6 months, and eventually revisited our clinic complaining of bilateral ear fullness, tongue numbness, and facial weakness that developed 2 days prior to the visit. She stated that the tube had been removed after symptom resolution. However, ear fullness recurred 1 month after removal and she received no additional treatment. Tympanic membranes were bulging bilaterally (Fig. 1A). She had right-sided facial weakness of grade II, according to the House-Brackmann classification [4]. Air-bone gaps of 35 dB were observed by PTA. Myringotomy and ventilation tube insertion were performed bilaterally to relieve symptoms. Yellowish granulation tissue exuded during myringotomy and was submitted for tissue culture. High dose steroids (60 mg prednisolone daily for 5 days and followed by tapering) were prescribed under the diagnosis of facial paralysis due to the aggravation of otitis media.

One week after initiating steroid therapy, all symptoms resolved. Facial symmetry was restored and tympanic membranes appeared to be normal with the tubes inserted (Fig. 1B). No microorganism growth was observed in tissue culture. However, 2 weeks after completing steroid treatment, ear fullness redeveloped and ventilation tubes were extruded with the granulation-like material. A tissue biopsy then revealed accumulations of eosinophils

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https://doi.org/10.1016/j.anorl.2018.03.009
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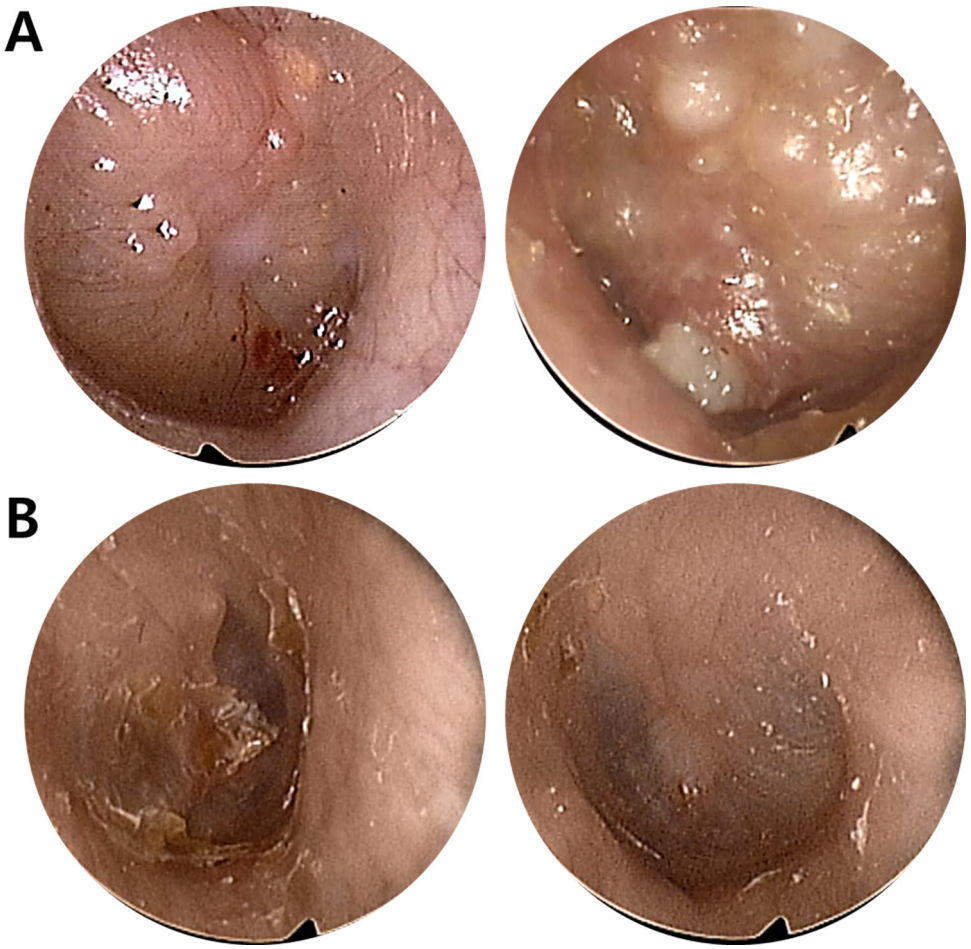

Fig. 1. Endoscopic findings of tympanic membranes. Tympanic membranes bulged bilaterally at the initial visit (A) but normalized after steroid treatment (B).

and Charcot-Leyden crystals (Fig. 2). Intratympanic steroid injection and oral steroids were then administered under the diagnosis of eosinophilic otitis media. Laboratory tests were performed and extremely high immunoglobulin (Ig) E and eosinophil counts (Ig E total: 1998 KU/L, eosinophil count: 1510/μL, serum eosinophils: 20.1%, eosinophil cationic protein [ECP]: 126.0/μg) were found. The patient continued to receive intratympanic steroid injection on a regular basis for 6 months and systemic steroids were administered when symptoms were aggravated. Her symptoms tended to respond well to only systemic steroids during follow-up. After 5 months of this treatment, she suddenly developed an itching sense of eyes and skin, and dyspnea with nocturnal aggravation. We then repeated the laboratory tests, which continued to reveal elevated Ig E (1706 KU/L) and eosinophil count (3900/μL) (serum eosinophils: 39.3%, ECP: 75.5/μg). The patient was then referred to the department of allergy and general workup was performed for HES. However, skin prick test, pulmonary function test, provocation test, echocardiography, parasite lab results from peripheral blood cell smear and abdominal computed tomography findings were normal. High dose systemic steroids (100 mg prednisolone daily) were administered for 5 days under the diagnosis of idiopathic HES. After systemic steroid treatment, all symptoms including ear fullness improved, tympanic membranes appeared normal, and air-bone gaps by PTA almost disappeared.

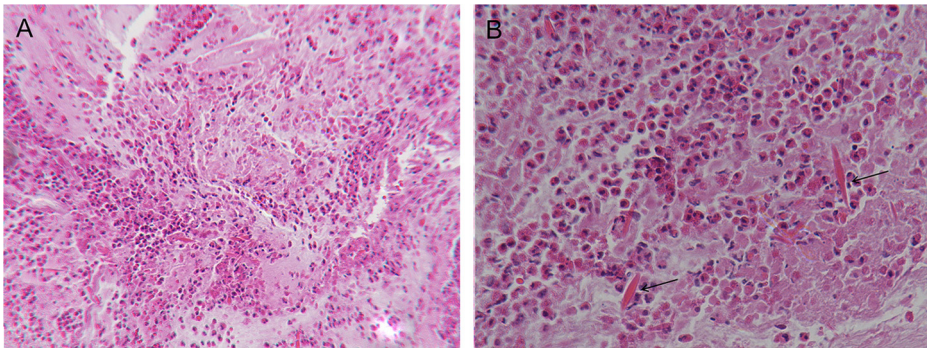

Fig. 2. Middle ear granulation biopsy showed inflamed granulation tissue with mucinous material and numerous eosinophils (A, H&E, × 200) and occasional Charcot-Leyden crystals (arrows) (B, H&E, × 400) produced by eosinophil breakdown.

At the visit 6 months after HES was diagnosed, she did not report any symptom. Oral steroids continue to be titrated to achieve effective symptom relief and a normal eosinophil count.

3. Discussion

Idiopathic HES is diagnosed when a patient has persistent eosinophilia more than 1500/μL recorded on at least two examinations with a minimum time interval of 4 weeks and organ dysfunction without a secondary cause of eosinophilia [1,5]. It mainly involves cardiovascular system, lung, skin, kidney, muscle and nervous system [2,5]. When HES involves the middle ear cavity, granulation tissue without calcification is usually found, and it can cause ear fullness and hearing loss [3]. Ventilation tube insertion and myringotomy are less effective on these obstructive symptoms, whereas systemic treatments, such as intraoral or intravenous steroid administration are effective [3]. When eosinophilic otitis media is aggravated, multiple polyps can be found in the middle ear cavity and symptom severity may be related to serum Ig E elevation [6,7]. In our patient, serum Ig E decreased when symptoms improved. Treatment includes local steroid administration, such as intratympanic steroid injection, and systemic steroid administration [8], although Ig E target therapy using omalizumab was recently reported [9].

Eosinophilic otitis media is characterized by resistance to conventional treatment for otitis media and highly viscous eosinophil-dominant effusion [8], and should be differentiated from HES. Symptoms and treatment for middle ear involvement are similar, but differential diagnosis warrants caution due to possibility of persistent hypereosinophilia and damage to other major organs. Churg-Struss syndrome, a rare systemic disease primarily characterized by hypereosinophilia, asthma and vasculitis, is also a consideration during the differential diagnosis of eosinophilic otitis media [8].

Patients with HES usually present with more general symptoms and often visit their internist. Middle ear effusion or hearing loss is not a typical clinical manifestation, and thus, it is uncommon that patients initially present to otolaryngologists. Furthermore, laboratory tests can be easily overlooked or missed, and as a result, diagnosis and treatment can be delayed.

When a patient is diagnosed with eosinophilic otitis media, careful attention should be made to non-specific skin symptoms or numbness and response to local treatment or systemic steroids should be evaluated. In such cases, further evaluation including laboratory tests, should be considered to exclude HES.

Two cases of HES involving the middle ear have been previously reported. The first case is a 39-year-old female, who presented with a non-productive cough and bilateral hearing loss, was found to have HES involving lung and middle ear. The second case is a 42-year-old female, who presented with a bilateral hearing loss, nasal obstruction, and dyspnea, and was diagnosed after eosinophilic pneumonia and eosinophilic otitis media had been confirmed by lung and middle ear biopsy. Both of these previously reported patients were managed using systemic steroids [2,5].

In our case, ventilation tubes were inserted at the initial visit under suspicion of middle ear effusion, but tubes were extruded prematurely and repeated insertions were ineffective due to the high viscosity of the exudate. Tissue biopsy and culture were performed under suspicion of eosinophilic otitis media, and steroids were administered orally and by intratympanic injection. When symptoms improved, oral steroids were discontinued and intratympanic steroid injections were maintained. However, symptoms then aggravated, and we became aware of the significance of previous laboratory results. As a result, a final diagnosis HES was delayed due to the initial misdiagnosis of eosinophilic otitis media. The bone conduction threshold of our patient was intact at last follow-up. However, progressive damage of the involved organ in HES can result in sensorineural hearing loss. We suggest that clinicians search for any impairment of inner ear function during the disease course of HES.

4. Conclusion

Eosinophilic otitis media and HES involving the middle ear share many clinical manifestations induced by the high viscosity of the exudate in the middle ear cavity. Furthermore, routine middle ear effusion therapy has little effect on both diseases. Local steroid administration is effective in eosinophilic otitis media, but not in HES, as occurred in our case. Prompt and accurate differential diagnosis is required for these diseases to ameliorate symptoms and promote recovery.

Disclosure of interest

The authors declare that they have no competing interest.

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