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CASE REPORT

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Atypical Ramsay Hunt syndrome (zoster sine herpete) with otitis media

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Abstract

A 21-year-old man presented with an acute onset of bilateral throbbing headache, left ear pain, tinnitus, and fever. There was no skin rash on his face. Otoscopy revealed hyperemia and exudate over the left tympanic membrane. The swab culture of the exudate grew methicillin-sensitive *Staphylococcus aureus*, and the patient was diagnosed as acute otitis media. Hearing loss and ipsilateral facial paralysis developed on hospital day 4. Despite the absence of typical bullous lesions, serology testing and polymerase chain reaction of the otic exudate for varicella-zoster virus were positive. The patient was finally diagnosed as zoster sine herpete.

KEYWORDS

acute otitis media, facial paralysis, varicella-zoster virus, zoster sine herpete

1 | INTRODUCTION

Zoster sine herpete (ZSH), classically defined as unilateral segmental radicular pain in the absence of an antecedent rash, has now become a distinct entity with a spectrum wider than what was originally described, with polymerase chain reaction (PCR) and serology testing providing definitive virologic confirmation of varicella-zoster virus (VZV) infection.[1] VZV reactivation without rash can present differently, including vasculopathy, radiculopathy, ophthalmic neuralgia, acute labyrinthitis, vagus nerve palsy, and facial nerve palsy.[2] ZSH has been reported in 8%-25% of patients with acute peripheral facial nerve palsy.[3] We report a case of facial nerve palsy due to ZSH while treating otitis media whose diagnostic clincher was the PCR of the otic exudate for VZV.

2 | CASE PRESENTATION

A previously healthy 21-year-old man presented to our emergency department with an acute onset of bilateral throbbing headache, left ear pain, and tinnitus for 4 days, followed by fever and fatigue. There was no nuchal rigidity, jolt accentuation, allodynia, or neuralgia. On admission, otoscopy revealed hyperemia and exudate over the left tympanic membrane with mild bulging, suggesting acute otitis media. His white blood cell count was 6.3×10^3 cells/mm^3, and C-reactive protein was <0.04 mg/dL. He was initially treated with intravenous ampicillin/sulbactam 3 g six-hourly. On day 3, the swab culture of the exudate grew methicillin-sensitive *Staphylococcus aureus*, and we switched his treatment to oral cefaclor 500 mg thrice daily. Although his fever subsided and headache improved gradually, otalgia, tinnitus, and dizziness

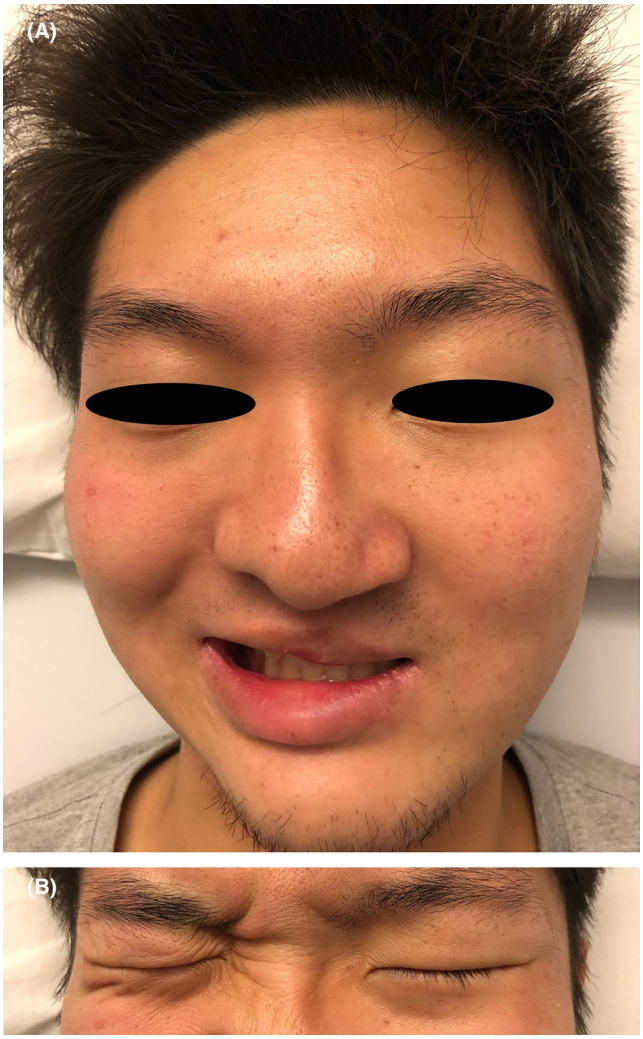

FIGURE 1 A, Facial palsy of the left side with the inability to lift the upper lip. B, Facial palsy of the left side with the inability to close eye

persisted. On day 4, he developed dysgeusia and hearing loss, followed by ipsilateral facial paralysis on day 7 (Figure 1). A head computed tomography scan did not show any bone involvement in the auditory ossicles or mastoiditis. There were no vesicles on the left pinna or external auditory canal. Since the otomicroscopic examination revealed no evidence of otitis media exacerbation, we suspected Ramsay Hunt syndrome (RHS) with grade V/severe palsy (House-Brackmann scale)[4] and started him on oral valacyclovir 1000 mg thrice daily and oral prednisone 60 mg daily. Valacyclovir and tapered prednisone were continued for 1 week, and his ear pain, tinnitus, and dizziness subsided. Serology testing for VZV revealed an immunoglobulin M (IgM) of 1.12 and IgG > 120 (normal values: IgM < 0.8 and IgG < 2.0), and the polymerase chain reaction (PCR) of the otic exudate for VZV was positive. An antigen/antibody test for human immunodeficiency virus was negative. During the treatment, no vesicle was observed in the auditory canal or on the auricle. These findings were consistent with the diagnosis of ZSH. Despite prompt treatment, grade III (moderate) facial palsy persisted, four weeks following discharge.

3 | DISCUSSION

RHS typically presents with a triad of ipsilateral facial paralysis, ear pain, and vesicles in the auditory canal or on the auricle.[5] Some cases with RHS exhibit facial paralysis with the absence of skin lesions, which is considered as a variety of ZSH and VZV infection in Bell's palsy.[2,6] ZSH is commonly identified as a localized radicular pain with virologic confirmation of VZV infection. ZSH was originally reported by the presence of VZV DNA in the cerebrospinal fluid of two patients with chronic radicular pain in the absence of rash that responded clinically and virologically to antiviral therapy.[7] VZV reactivation without rash can present differently, including vasculopathy, radiculopathy, ophthalmic neuralgia, acute labyrinthitis, vagus nerve palsy, and facial nerve palsy.[2] Diagnosis of ZSH was complicated in this case since otitis media can also cause facial palsy and otalgia. DNA-PCR of the ear exudate and serologic testing aided in the diagnosis. One case report showed that PCR analysis of VZV DNA in auricular skin exudates was a useful diagnostic tool for the diagnosis of ZSH.[8]

Zoster sine herpete can be confirmed by either a fourfold increase in VZV antibody titers or the detection of VZV DNA in the skin, blood mononuclear cells, saliva, cerebrospinal fluid, or middle ear fluid.[6] Furuta reported that detection of VZV DNA in oropharyngeal swabs by PCR was more useful than serological assays for the early diagnosis of ZSH in patients with acute peripheral facial nerve palsy.[3] To our best knowledge, few clinical research to evaluate the sensitivity and specificity of DNA-PCR of the ear exudate and serologic testing for the diagnosis of ZSH has been reported, so further studies are needed.

Why should clinicians confirm VZV infection in Bell's palsy? HSV activation is widely accepted as the probable cause of Bell's palsy, which is the most common cause of acute peripheral facial palsy. VZV is the second most common virus implicated in Bell's palsy. Peitersen identified VZV infection in 116 (6.8%) patients among 1701 cases of Bell's palsy.[9] The neurological recovery rate in Bell's palsy patients with VZV reactivation was lower than that in patients with HSV-1 reactivation.[10] This suggests that confirming VZV infection in Bell's palsy may help in assessing the prognosis.

Learning point from this patient is that in cases involving facial paralysis and ear pain, clinicians should consider a diagnosis of ZSH and PCR testing of the ear exudate, even in the absence of typical bullous lesions.

4 | CONCLUSION

We report a case of ZSH while treating otitis media. Although otitis media itself can cause facial nerve palsy and ear pain, complicating the diagnosis of ZSH, DNA-PCR for VZV of the ear exudate was useful for the detection and the serologic reactivation of VZV. ZSH should be considered even in the absence of typical bullous lesions, when ear pain persists and facial paralysis occurs during otitis media treatment.

CONFLICT OF INTEREST

The authors have stated explicitly that there are no conflicts of interest in connection with this article.

CONSENT FOR CASE REPORT

Patient consent has been obtained.

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