AnnalsofClinicalandMedical Case Reports

CaseReport ISSN2639-8109Volume9

RamsayHuntSyndromewithChickenpox:ACaseReportinElderWoman

MaMX,ZhangXH,LiYQ,WuSQ,LangN*,Jian-XunRen*

Xiyuan Hospital of ChinaAcademy of Chinese Medical Sciences, China

***Correspondingauthor:**

Na Lang,
DepartmentofDermatology,XiyuanHospitalofChina Academy of Chinese Medical Sciences, BeiJing, China, Tel: +86-628352089;
E-mail:langna96@126.com

Jian-Xun Ren,
Experimentalcenter,XiyuanHospitalofChina Academy of Chinese Medical Sciences, BeiJing, China,Tel:+86-62835608;E-mail:rjx131@sina.com

Received: 05 Oct 2022
Accepted: 17 Oct 2022
Published:21Oct2022
JShort Name:ACMCR

Citation:

Jian-Xun Ren, Ramsay Hunt Syndrome with Chickenpox:ACaseReportinElderWoman.AnnClinMedCase Rep. 2022; V10(3): 1-4

Keywords:

RamsayHuntsyndrome;Herpeszoster;Varicella;Chickenpox

1. Abstract

Ramsay Hunt syndrome (RHS) is an infectious disease characterized by delayed activation of latent herpes zoster virus in the geniculate ganglion and subsequent spread to cranial nerve. Although herpesvirus can also cause chickenpox and herpes zoster, thedelayedchickenpoxisararepresentationofRHS.Wepresent the case of a 76-year-old female patient withAlzheimer's disease appearedunstablegaitfor2days.Thepatienthadleftfacialpalsy with concomitant skin lesions in the left external auditory canal. After exclusion of ischemic cerebrovascular diseases, the patient wasdiagnosedwithRHSbytypicalsymptoms.Subsequently,the typicalchickenpoxbrokeoutontherightauricleduringhospitalization.Afterantiviralandhormonetherapy,thepatient'sRHS-associatedsymptomswererelieved.EarlydiagnosisofRHS,andthe use of acyclovir-corticosteroid to relieve inflammation and injury of nerves can bring a crucial effect on prognosis of cranial nerve damage.

2. Introduction

RamsayHuntsyndrome(RHS)isacomplicationofvaricella-zos- ter virus infection with involvement of the seventh and eighth cranial nerves. The clinical symptoms are characterized by acute facialpalsyandvestibulocochlearinjurywithaherpeseruptionon the external auricular [1]. RHS was first described by J. Ramsay Hunt in 1907. RHS is a special form of herpes zoster caused by reactivationandreplicationofvaricellazostervirus(VZV)inthe geniculate ganglion of the facial nerve, and less than 1% of zoster cases involve the facial nerve result as RHS [2].VZVbelongs to human herpes viruses α subfamily, which are double-stranded DNAvirusesthattransmissibleviarespiratorydroplets.Afterprimaryvaricellainfection,theVZVpersistsinthespinalandcranial nervegangliaoverlifetime,andtheoverwhelmingmajorityofinfectedindividualshavenosymptoms.ReactivationoflatentVZV is triggered at a later stage in a state of compromised immune to present as herpes zoster.Although RHS is frequent in adults and increases with age due to primary exposure to the virus in childhood, the disease is rare in older individuals.

After reactivation and replication, the viruses travel through the sensory fibers of facial nerve into the dermatome associated with the involved ganglion around the auricular and external auditory meatus, which induces the typical clinical characteristics of pain and rash in herpetiform distribution. Beyond the dermatological manifestations,reactivatedVZValsostimulatetheadjacentmotor branches of the facial nerve, which leads to facial muscle pare-sis and balance disorders, as well as lacrimal and nasal secretion. SomepeopleinfectedwithVZVcancausechickenpoxatthetime of viruses first attack, however, VZV reactivation can produce a variety of chronically neurological damage with or without rash on the ear or in the mouth, and rarely, with chickenpox. The diagnosis is mainly based on the typical clinical manifestations.Theappropriatecorticosteroidsandantiviraltherapyr

esultedina

good prognosis. But there is lack of data on long-term outcomes. Here, we report the case of an elder patient suffered from RHS with chickenpox, which shows the early diagnostic challenges of this syndrome.

The early diagnosis of Ramsay Hunt syndrome is essential, as prognosis of cranial nerve damage depends on the time at which corticosteroid-based therapy is started.

3. Case Report

A76-year-oldfemalepatienthadexhibitedunsteadygaitfor2days with left-sided facial weakness. She reported no vertigo, tinnitus, nausea, and vomiting. One week ago, before she presented to our hospital, she was found rush appeared on her left ear.The patient had suffered fromAlzheimer's disease for more than 5 years and prescribed memantine hydrochloride tablets (10 mg QD) and donepezil hydrochloride tablets (10 mg QN) for the past year. She hadnohistoryofunderlyingdiseases.Tofurtherconfirmwhether the cerebral disease existed, she took Cranial CT and MRI which revealed no obvious abnormalities.

Physical examination revealed a drunken gait and facial paralysis of the left side was found. The facial voluntary movement was evaluated with the House-Brackmann grading system, and the peripheral facial paralysis was belonged to be House-Brackmann grade III. There are many purulent secretions in the left ear (Figure1),however,norashwasobservedontherightear,trunk,and limbs. The patient had hearing loss and was unable to cooperate with pure tone audiometry. We also found that the left forehead wrinkles disappeared, the left nasolabial groove became shallow in this patient (Figure 2).

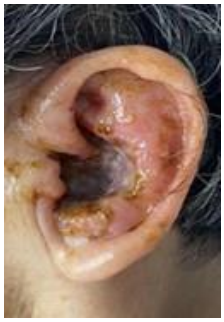

Figure 1: Left ear of the patient: white pus with paste of Chinese medicine (yellow part).

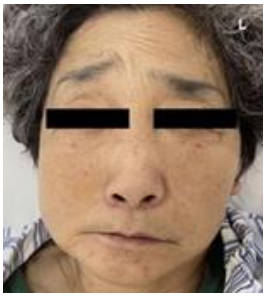

Figure2: Thepatient'sleftfacialmusclewasweakandappearedflaccid.

Basedontheclinicalsignsandsymptomsthatmainlymanifestshe wasdiagnosedtheRHS.Antiviraltherapywithfamciclovirtablets and prednisone acetate tablets was begun at a dosage of 0.75 g/d and 10 mg/d, respectively.

During admission, new herpes focus appeared on the patient's trunk. On examination, red herpes, as a typical chickenpox were seenintheskinofthechestandback.Theherpesblisterswerethe sizeofsoybeanswithuniformshape,andtherewasnohaloaround them. The blister wall was thin, and the blister fluid was clear. The herpes itched, some of them were scratched up and ulcerated (Figure 3).

After 3 days of treatment, the patient was able to walk normally and there was still some new chickenpox left on the trunk. Since hermaincomplaintwasresolved,shewasdischargedfromhospital,andadvisedtomaintainthecurrenttreatmentandtobevigilant about protection in others.

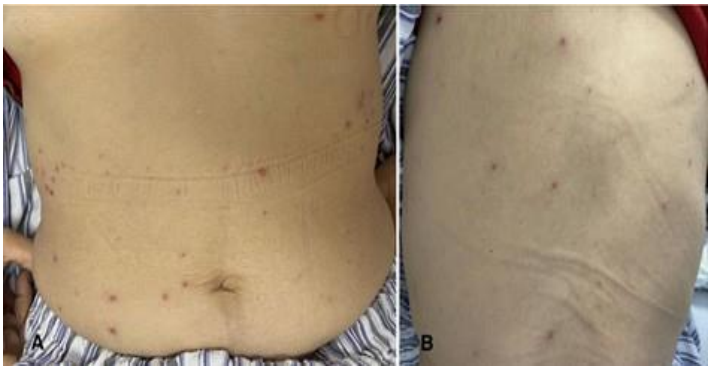

Figure3: Thepatient'strunkwascoveredwithsmallblisters,andsomeof the blisters had been broken.

4. Discussion

Acute facial paralysis in elder can be caused by a range of disorders, including ischemic cerebrovascular diseases, trauma, infection, metabolic and neoplastic factors. In elderly patients with sudden gait unsteadiness and facial paralysis, acute cerebral infarctionismorepronetobeconsideredandthepossibilityofRHS couldbeignored.RHSusuallyisdiagnosedonaclinicalbasis,as laboratory testing is often slow or impractical. In definition, RHS is characterized by combination of acute facial nerve palsy and vesiculareruptionoftheskinofpinnaandexternalauditorycanal caused by VZV. VZV is present worldwide and 98% of the adult population in the United States is seropositive for VZV [3]. Two clinicalstudiesdemonstratedthatRHSwasthecauseoffacialpal- sy in 16.7% of children, and 18.1% of adult [4-5], respectively. TheprevalenceoffacialparalysisinducedbyVZVreactivationin adult was significantly higher than children, which reflect an underlyingimmunocompromisedstatuswithadvancedage.Multiple cranialnerveganglia,includinggeniculateganglionandperipheralgangliaofcranialnervesVIII,IXandXinvolvementfrequently occurred in RHS [6]. Pathophysiological characteristics of multiplecranialnerveinvolvementassociatedwithRHSstillremains

unclear. Hunt et al suggested that adjacent gasserian, petrous, accessory, jugular, plexiform, the second, and the third cervical dorsal root ganglia may form chain allowing the extension of the ganglionitis[7].TheperineuralspreadofthereactivatedVZVcan travels through the small branches of the infected carotid artery, middle meningeal artery, and ascending pharyngeal artery that supply blood to cranial nerves V, VII, IX, X, XI and XII cranial nerves. In this patient,VZVhad gradual progressive involvement of lower cranial nerves VII, VIII, IX and X associated with herpetic eruptions on the outer aspect of the ear. Thus, the combinationdysfunctionofmotorandsensoryinvolvementleadstomotor weakness, muscular spasm or in the form of concomitant motor movements. Clinical symptoms presented in this case as balance dysfunction, which is typical symptoms in RHS and blame to the cochlear nerve lesions [8]. The geniculate ganglion sits between the cochlea and the tympanic cavity, there are many reasons to explain how VZV infect the cochlear nerve [9].

Firstly, the vestibular nerve and can be transmitted directly throughneurons,previouslystudieshaveconfirmedthatVZVcan betransmittedthroughaxons[10].Besides,VZVcanbetransmittedthroughtheblood,Topographically,thegeniculateganglionis nearbycochlearnerveandoriginfromtheposteriorcirculationso that VZV can travel from the geniculate ganglion to the cochlear nerve.Another important point is thatVZVwas also found in the spirochetes and vestibular ganglion of some individuals without clinical manifestations in early studies [11], which means virus could latent directly in the vestibular ganglion.

Up to now, the strict definition of the RHS is peripheral facial nerve palsy accompanied by an erythematous vesicular rash on theear(zosteroticus).ButSomepatientsdevelopperipheralfacial paralysis without rash which is really difficult to distinguish clinicallyfromBellpalsy.Murakamietal.identifiedRHSzostersine herpete in six (19%) of the patients in a study of 32 patients with isolated peripheral facial palsy [12].

RHShasanincidenceofabout5per100,000peopleperyear[13], while RHS combined with chickenpox is quite rare for most her- pes patients. During an episode of zoster, vesicular rashes tend to appear within a single dermatome [14]. In this case, the patient varicella spread over trunk, displayedVZVinfected multiple spinal ganglia.

VZV infection causes primary varicella (chickenpox). Varicel-lazostervirusishighlycontagiousandistransmittedthroughairbornedropletsordirectcontactwithvesicularfluid.Afterprimary chickenpox,chickenpoxisusuallyseeninsusceptibleunvaccinat- ed individuals, but can also present in individuals who had been previously vaccinated [15]. Therefore, the susceptible immunocompromise is a key factor in RHS with chickenpox. Although VZVasaDNAvirusismoreconservativethanRNAvirus,many mutations of VZV have still found [16]. The gene fragment encodingglycoproteinEcouldhaveabasemutationatasinglesite, which resulted to amino acid changes. After mutation, the infectivity of VZV may change, and the infected strain of this patient may be mutated [17]. Acyclovir is the first-line treatment option forRHStopreventdiseaseprogressionregardlessofimmunesta- tus or disease severity.

5. Conclusion

This case demonstrates an elder RHS patient which means VZV had infected and reactivated, it is possible that chickenpox presentation is now so uncommon in clinical practice that it is more difficult to identify. Complications of chickenpox can present in immunodeficient individuals, or in healthy adults. Advanced age can be an important factor to chickenpox, especially in high-risk individuals. Early recognition of RHS with chickenpox is also important to prevent transmission to others, especially to those at increased risk of complications, including susceptible adults, and susceptible immunocompromised individuals.

References

1. Khan YMT, Fatema N. Ramsay Hunt syndrome. Pan Afr Med J. 2019; 34: 201.
2. Sommer T, Karsy M, Driscoll MJ, et al. Varicella-Zoster Virus Infection and Osteomyelitis of the Skull. World Neurosurg. 2018; 115: 297-300.
3. Schmid DS, Jumaan AO. Impact of varicella vaccine on varicella-zoster virus dynamics. Clin Microbiol Rev. 2010; 23(1): 202-17.
4. Yeo SW, Lee DH, Jun BC, et al. Analysis of prognostic factors in Bell'spalsyandRamsayHuntsyndrome.AurisNasusLarynx.2007; 34(2): 159-64.
5. HatoN,KisakiH,HondaN,GyoK,etal.RamsayHuntsyndromein children. Ann Neurol. 2000; 48(2): 254-6.
6. NonnekesJ,RůžičkaE,SerranováT,etal.Functionalgaitdisorders: Asign-basedapproach.Neurology.2020;94(24):1093-1099.
7. Kuhweide R, Van de Steene V, Vlaminck S, et al. Ramsay Hunt syndrome: pathophysiology of cochleovestibular symptoms. J Laryngol Otol. 2002; 116(10): 844-8.
8. TsauPW,LiaoMF,HsuJL,etal.ClinicalPresentationsandOutcome Studies of Cranial Nerve Involvement in Herpes Zoster Infection:A Retrospective Single-CenterAnalysis. J Clin Med. 2020; 9(4): 946.
9. de Castro DC, Marrone LC. Neuroanatomy, Geniculate Ganglion. 2021.
10. Mayberg M, Langer RS, Zervas NT, et al. Perivascular meningeal projections from cat trigeminal ganglia: possible pathway for vascular headaches in man[J]. Science. 1981; 213: 228-230.
11. Furuta Y, Takasu T, Suzuki S, et al. Detection of latent varicella-zoster virus infection in human vestibular and spiral ganglia. J Med Virol. 1997; 51: 214-16.
12. Murakami S, Honda N, Mizobuchi M, et al. Rapid diagnosis of varicellazostervirusinfectioninacutefacialpalsy,Neurology.1998; 51(4): 1202–1205.
13. CrouchAE,HohmanMH,AndaloroC.RamsayHuntSyndrome.

2022May1.In:StatPearls[Internet].TreasureIsland(FL):StatPearls. 2022.

14. Crouch AE,HohmanMH, AndaloroC.RamsayHuntSyndrome. 2022.
15. AndreiG,SnoeckR.AdvancesandPerspectivesintheManagement ofVaricella-ZosterVirus Infections. Molecules. 2021; 26(4): 1132.
16. KennedyPGE,MogensenTH,CohrsRJ.RecentIssuesinVaricella-Zoster Virus Latency. Viruses. 2021; 13(10): 2018.
17. HarbeckeR,CohenJI,OxmanMN.HerpesZosterVaccines.JInfect Dis. 2021; 224(12 Suppl 2): S429-S442.