Received: 2017.08.26
Accepted: 2017.10.16
Published: 2018.01.18

e-ISSN 1941-5923
© Am J Case Rep, 2018; 19: 68-71
DOI: 10.12659/AJCR.906834

Ramsay Hunt Syndrome with Multiple Cranial Neuropathy in an Human Immunodeficiency Virus (HIV) Patient

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

EF 1 **Divya Arya**
EF 2 **Tushar Bajaj**
F 3 **Jose Gonzalez**
E 4 **Rene Elkin**

1 Department of Internal Medicine, Bronx-Lebanon Hospital, Bronx, NY, U.S.A.
2 Department of Medicine, American University of the Caribbean, Cupecoy, St. Maarten
3 Department of Internal Medicine, Bronx-Lebanon Hospital, Bronx, NY, U.S.A.
4 Department of Neurology, Bronx-Lebanon Hospital, Bronx, NY, U.S.A.

Corresponding Author: Divya Arya, e-mail: divya.arya90@gmail.com
Conflict of interest: None declared

Patient: Male, 29
Final Diagnosis: Ramsay Hunt syndrome with multiple cranial nerve involvement
Symptoms: Chest pain • dizziness • dysesthesia • dysphagia • ear pain • facial palsy • gait imbalance • headache • vertigo
Medication: Emtricitabine/tenofovir alafenamide and dolutegravir
Clinical Procedure: —
Specialty: Neurology

Objective: Unusual clinical course
Background: Ramsay Hunt syndrome is a rare otologic complication resulting from varicella zoster virus reactivation that can present with a myriad of clinical presentations. Most common being triad of ear pain, vesicles at auricle, and ear canal with same side facial palsy.
Case Report: We report a case of a 29-year-old male with a human immunodeficiency virus (HIV) infection who presented with left facial palsy, vesicles, pain in the left ear, dysphagia, dizziness, and headache resulting from multiple cranial nerves involvement such as cranial nerve V, VII, VIII, IX, and X.
Conclusions: This case report raises awareness among general practitioners to investigate for Ramsay Hunt syndrome in HIV patients presenting with ear pain with a thorough neurological exam and emphasize on the interplay of different specialties in managing these patients.

MeSH Keywords: Bell Palsy • Deglutition Disorders • Herpes Zoster Oticus • HIV

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/906834>

1240 1 3 29

Background

Various terms have been used in literature to define the vivid presentation of Ramsay Hunt syndrome including herpes zoster oticus, herpes zoster cephalicus, and zoster sine herpete. Latent varicella zoster virus reactivation in the ganglion causes a sensory and motor neuropathy. Cranial nerves V, VII, VIII, IX, X, XI, and XII could be involved with variable frequencies [1–5]. There is also an anecdotal report of herpes simplex type 2 virus (HSV 2) causing Ramsay Hunt syndrome [6]. Multiple risk factors have been identified which predisposes individual to this disease. The diagnosis is based on the classical triad of ear pain, vesicles and ipsilateral facial paralysis. Other manifestations, which have been observed include dysgeusia, hearing impairment, tinnitus, hyperacusis, vertigo, changes in lacrimation, and hoarseness of voice [1,2]. Combination of antiviral and corticosteroids are the mainstay of treatment along with other supplementary modalities such as artificial tears, eye patch, and facial exercises [7]. This case report focuses on how a simple clinical presentation of ear pain in patients with known risk factors like immunocompromised state can lead to diagnosis of Ramsay Hunt syndrome.

Case Report

A 29-year-old HIV positive male African American presented to the emergency department with left-sided facial palsy. Two weeks prior to this, he developed bilateral ear pain and was treated for an ear infection with antibiotics at another hospital. However, even after completing the course of antibiotics, the ear pain did not resolve. On the day of admission, the patient woke up with dysesthesia of the left-sided tongue, left-sided facial palsy, headache, dizziness defined as gait imbalance, and chest pain.

His other medical conditions include human immunodeficiency virus (HIV) (he was on emtricitabine/tenofovir alafenamide and dolutegravir- noncompliant), hypertension, asthma, major depressive disorder and chickenpox as a child. He admitted using recreational drugs and smoking tobacco.

The patient was admitted to our general medical floor and placed on airborne and contact isolation. On initial evaluation, his vital signs were in the optimal range. On physical examination, he was agitated, his left ear was erythematous, edematous, and vesicles were seen in the external auditory canal, with no lesions seen in the oral cavity. On neurological examination, left-sided lower motor neuron complete facial palsy was apparent – inability to close left eye (Figure 1), inability to smile uniformly (Figure 2) and inability to frown the left forehead. The facial function was graded as IV on House-Brackman Classification. This presentation was associated with decreased

Figure 1. Patient instructed to close both eyes. Unable to do on left side.

Figure 2. Patient instructed to smile.

sensation to touch and temperature on left side of the face, unstable gait, and inability to swallow properly.

Pertinent laboratory values included – white blood cell count of 7.3 k/uL, hematocrit of 41, platelet count of 260 k/uL, and blood urea nitrogen/creatinine ratio of 10/1.0. Serum electrolytes, liver enzymes, coagulation profile, and cardiac markers were within normal limits. Urine toxicology was positive for cocaine. His CD4 count was 348 cells/mm³, and computer tomography of the head without contrast showed no abnormalities. Chest x-ray did not show any anatomical abnormality.

A clinical diagnosis of Ramsay Hunt syndrome was made. An infectious disease and neurology evaluation was requested. The patient was started on valacyclovir 1,000 mg three times a day, prednisone 90 mg once a day for a total of five days, artificial tears and analgesics were given as needed. The patient was started on puree and thin liquid diet due to swallowing problems with aspiration precautions.

On day 5, the patient showed improvement in swallowing abilities and gait but with no change in the facial palsy. The patient gradually improved, and he was able to safely swallow and placed on regular diet by the swallow team. Physical and occupation therapy evaluations were done, and appropriate rehabilitation measures were initiated. He was discharge in stable condition and advised to follow-up in ambulatory clinics for further management. However, the patient did not follow-up in clinics, but we were able to contact him by phone, and further minimal improvement was thus noted.

Table 1. Recovery rate seen in cases described by Shim et al. with and without multiple cranial nerve involvement.

	With multiple cranial neuropathy	Without multiple cranial nerve neuropathy
Recovery rate	54.5	82.9
Complete recovery rate (%)	27.3	67.7

Discussion

Ramsay Hunt syndrome describes the classical presentation of ear pain, vesicles, and facial paralysis resulting after a latent virus reactivation in the geniculate ganglion. Due to its variable clinical presentations, different terms have been used to describe this syndrome such as herpes zoster oticus (J. Ramsay Hunt), herpes zoster cephalicus (Blakley et al.) [8], and zoster sine herpete (without skin eruptions [1,9–12]). Testing via polymerase chain reaction (PCR) is not recommended as it results in no change in management [13].

Based on the clinical presentation of our case, the most appropriate descriptive term would be herpes zoster cephalicus as there was involvement of cranial nerve V (affecting the sensation of the left face and swallowing), cranial nerve VII involvement (facial palsy and swallowing), cranial nerve VIII (vertigo and gait imbalance), and cranial nerves IX and X (dysphagia).

There are numerous risk factors, which have been reported in the literature, that result in reactivation of the latent virus such as aging, emotional stress, smoking, diabetes, depression, immunocompromised state, cancer, organ transplantation, and certain medications including but not limited to corticosteroids and immunosuppressive agents [5,14–20]. Our patient was HIV-positive with a history of non-compliance with medications, suffered from depression, and had a tobacco smoking history.

Herpes zoster incidence is 15 times more likely in HIV-infected individuals compared to their age-matched controls [21]. The immunocompromised state of HIV patients subjects them to disseminated infection, neurological complications, post-therapeutic neuralgia, and recurrent infections [5]. CD4+ cell count ($\leq 200/\text{mm}^3$) further adds up to patient vulnerability.

The hypothesis for involvement of multiple cranial nerves encompasses factors like anatomical proximity, embryological origin of the nerves, hematogenous spread, and transaxonal spread [22–25]. In our case, the patient had dysphagia and dizziness which improved to a large extent; however, facial palsy remained unchanged. This could be due to the spread of less intense inflammation from effected ganglion, which resolves rapidly. Espay and Bull described a similar progression in an 83-year-old female with HIV-negative status who initially presented with hearing impairment, hoarseness, dysphagia, and left-sided facial palsy, who then received treatment with

Figure 3. Prognosis of different type of Bell's palsy.

acyclovir 800 mg, five times a day for 10 days, and showed moderate improvement in her ability to swallow; however, at her 120-day follow-up it was noted that other impairments continued [26]. Inclusion of multiple cranial nerves results in a more debilitating course, as was also the case with our patient. Shim et al. described 11 patient cases with multiple cranial nerve involvement as summarized in Table 1 [27]. Figure 3 describes the prognosis of facial palsy conditions [1,5,27].

Kinishi et al. found a significant p -value of 0.01 when a combination of methylprednisone and acyclovir was used rather than methylprednisone alone. Monsanto et al. provided a thorough review of the literature and found favorable outcomes with the this dual therapy [7,28]. In another review of the literature, surgical procedures such as microvascular decompression and rhizotomy as therapeutic interventions have been described [29].

Conclusions

Ramsay Hunt syndrome is a clinically diagnosed condition, which needs a very careful approach to achieve diagnosis and management as it affects an individual physically and psychologically. A proper neurological examination to define the extent of involvement is critical, as is starting steroid and antiviral therapy early for prevention of progression of disease. It is also important to have timely follow-up to monitor for signs of resolution or progression of disease.

Acknowledgements

We would like to acknowledge Dr. Manisha Singh, Dr. Suresh Nayudu the Associate Program Directors and Dr. Sridhar Chilimuri, Program Director of Bronx Lebanon Hospital Center.

Conflicts of interest

None.

References:

1. Adour KK: Otolological complications of herpes zoster. *Ann Neurol*, 1994; 35(Suppl.): S62-64
2. Mishell JH, Applebaum EL: Ramsay-Hunt syndrome in a patient with HIV infection. *Otolaryngol Head Neck Surg*, 1990; 102(2): 177-79
3. Aviel A, Marshak G: Ramsay-Hunt syndrome: A cranial polyneuropathy. *Am J Otolaryngol*, 1982; 3: 61-66
4. Payton RJ, Dawes JDK: Herpes zoster of the head and neck. *J Laryngol Otol*, 1972; 86: 1031-55
5. Macsumoto Y, Yanagihara N, Sdamoto M: Gd-DTPA imaging in peripheral facial palsy. *Facial N Res Jpn*, 1991; 11: 93-96
6. Diaz GA, Rakita RM, Koelle DM: A case of Ramsay Hunt-like syndrome caused by herpes simplex virus type 2. *Clin Infect Dis*, 2005; 40(10): 1545-47
7. Monsanto RD, Bittencourt AG, Bobato Neto NJ et al: Treatment and prognosis of facial palsy on Ramsay Hunt syndrome: Results based on a review of the literature. *Int Arch Otorhinolaryngol*, 2016; 20(4): 394-400
8. Blackley B, Friedmann I: Wright Herpes zoster auris associated with facial nerve palsy and auditory nerve symptoms. *Acta Otolaryngol*, 1967; 63: 533-50
9. Vafai A, Berger M: Zoster in patients infected with HIV: A review. *Am J Med Sci*, 2001; 321(6): 372-80
10. Gildeen DH, Dueland AN, Devlin ME et al: Varicellazoster virus reactivation without rash. *J Infect Dis*, 1992; 166: S30-34
11. Meylan PRA, Miklossy J, Iten A et al: Myelitis due to varicella-zoster virus in an immunocompromised patient without a cutaneous rash. *Clin Infect Dis*, 1995; 20: 206-8
12. Hunt JR: On herpetic inflammation of the geniculate ganglion and its complications. *J Nerv Ment Dis*, 1907; 34: 73-96
13. Furuta Y, Takasu T, Fukuda S et al: Detection of varicella-zoster virus DNA in human geniculate ganglia by polymerase chain reaction. *J Infect Dis*, 1992; 166(5): 1157-59
14. Straus SE, Oxman MN: Varicella and herpes zoster. In: Freedberg IM, Eisen AZ, Wolff K et al. (eds.), *Dermatology in general medicine*, 5th ed. Vol 2. New York: McGraw-Hill, 1999; 2427-50
15. Whitley RJ: Varicella-zoster virus. In: Mandell GL, Bennett JE, Dolin R (eds.), *Principles and practice of infectious diseases*, 4th ed. New York: Churchill Livingstone, 1995; 1345-51
16. Arvin AM: Varicella-zoster virus. *Clin Microbiol Rev*, 1996; 9: 361-81
17. Andiman RM, Brunton SA, Lawrence L et al: Herpes zoster: Presentations and pitfalls. *Patient Care*, 1994; 28: 2-11
18. Irwin M, Castlow C, Williams H et al: Cellular immunity to Varicella-Zoster virus in patients with major depression. *J Infect Dis*, 1998; 178S: 104-8
19. Guidetti D, Gabbi E, Motti L, Ferarini G: Neurological complications of herpes zoster. *Ital J Neurol Sci*, 1990; 11: 559-65
20. Irwin MR, Levin MJ, Carrillo C et al: Cellular immunity to Varicella-Zoster virus in patients with major depression. *Brain Behav Immun*, 2011; 25(4): 759-66
21. Buchbinder SP, Katz MH, Hessol NA et al: Herpes zoster and human immunodeficiency virus infection. *J Infect Dis*, 1992; 166(5): 1153-56
22. Hunt JR: Geniculate neuralgia (neuralgia of the nervus facialis): A further contribution to the sensory system of the facial nerve and its neuralgic conditions. *Arch Neurol Psychiatry*, 1937; 37: 253-85
23. Hunt JR: The symptom-complex of the acute posterior poliomyelitis of the geniculate, auditory, glossopharyngeal and pneumogastric ganglia. *Arch Intern Med*, 1910; 5: 631-75
24. Lapresle J, Lasjaunias P: Cranial nerve ischaemic arterial syndromes. *Brain*, 1986; 109: 207-15
25. Jonsson L, Tien R, Engström M et al: Gd-DPTA enhanced MRI in Bell's palsy and herpes zoster oticus: An overview and implications for future studies. *Acta Otolaryngol*, 1995; 115: 577-84
26. Espay AJ, Bull RL: Petrositis in Ramsay Hunt syndrome with multiple cranial neuropathies. *Arch Neurol*, 2005; 62(11): 1774-75
27. Shim HJ, Jung H, Park DC et al: Ramsay Hunt syndrome with multicranial nerve involvement. *Acta Otolaryngol*, 2011; 131(2): 210-15
28. Kinishi M, Amatsu M, Mohri M et al: Acyclovir improves recovery rate of facial nerve palsy in Ramsay Hunt syndrome. *Auris Nasus Larynx*, 2001; 28(3): 223-26
29. Rupa V, Saunders RL, Weider DJ: Geniculate neuralgia: The surgical management of primary otalgia. *J Neurosurg*, 1991; 75: 505-11