

A case of herpes zoster with cranial polyneuropathy involving multiple cranial nerves

H. Jaafoura, M. Tbini, E. Chebil, N. Ben Moussa, I. Riahi, R. Lahiani, S. Mannoubi, M. Ben Salah
 Department of Otorhinolaryngology-Head and Neck Surgery, Charles Nicolle Hospital, Tunis, Tunisia

Medicine faculty of Tunis. El Manar University

ABSTRACT

Introduction: In the head and neck region, the reactivation of varicella zona virus (VZV) affect frequently the VII and VIII cranial nerves (Ramsay Hunt syndrom) but rarely other cranial nerves may be involved.

Case report: We report the case of a 59 years-old man who presented to our department with hoarseness, vesicular eruption in the left cheek and the left ear, left side otalgia and dizziness. Based on physical examination, herpes zoster with cranial polyneuropathy involving cranial nerves: V, VII, VIII, IX, and X was suspected and the patient was treated by antivirals and corticosteroids. After re-education and speech therapy, he obtains a partial recovery.

Conclusion: Herpes zoster is a rare medical condition that should be evoked in case of unilateral or multiple cranial neuropathies regardless of whether skin or mucosal lesions exist or not. Treatment is based on antivirals and corticosteroids.

Key words: Herpes Zoster, Polyneuropathy, Trigeminal Nerve, Ramsay Hunt Syndrome, Glossopharyngeal nerve, Vagus Nerve.

INTRODUCTION

After a primary infection (chickenpox), the varicella zona virus (VZV) remains dormant in the ganglia of the spinal cord and cranial nerves and then becomes reactivated decades later. The reactivated VZV reaches the skin through axons usually causing vesicular eruption restricted to one or few dermatomes, pain in sensitive branches and palsy in motor ones. In the head and neck region, the most frequent presentation is Ramsay Hunt syndrome which involve the VII and VIII cranial nerves but rarely other cranial nerves may be involved.

CASE REPORT

We report the case of a 59 years-old man with no significant past medical history. He was complaining of a week history of sore throat, dysphagia, hoarseness and dyspnea. He consulted first a pulmonologist (respiratory physician) for his dyspnea, who performed a bronchoscopy revealing only a left vocal fold paralysis. In the meanwhile, the patient developed a vesicular eruption in the left cheek and the left ear, headache, left side otalgia and dizziness, so he was referred to our department.

Physical examination showed vesicles in the left auricle and external auditory canal (fig 1), and few vesicles at the ipsilateral cheek in the maxillary nerve "V2" area. Peripheral facial nerve palsy was noticed in the left side (fig 2). Gag reflex was absent and the uvula was deviated to the right. Flexible fiberoptic laryngoscopy revealed a paralysis of the right vocal fold fixed with mucosal lesions (fig 3). Ophthalmic examination revealed no pathological findings.

Laboratory tests were normal. Pure-tone audiometry showed a neurosensorial hearing loss of 40dB.

Videonystagmography (VNG) was performed and revealed a left side vestibular deficit.

A non-contrast computed tomography (CT) of the head and temporal bone revealed no abnormalities.



Figure 1 : Vesicular eruption in the left ear



Figure 2 : Left peripheral facial nerve palsy.

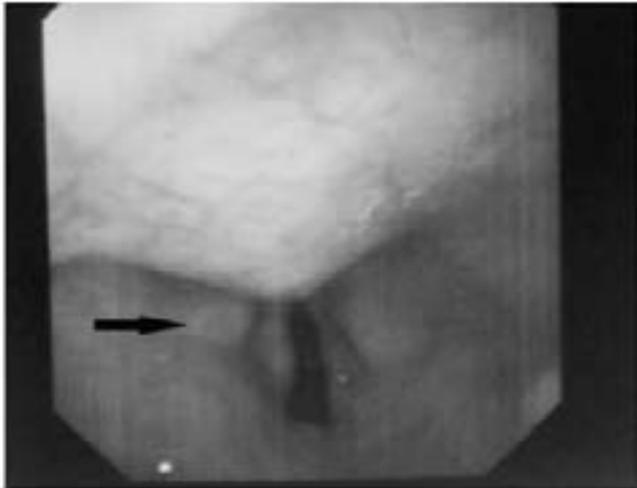


Figure 3 : Left vocal fold paresis with mucosal lesion

On the basis of this finding, a Ramsay Hunt syndrome associated with multiple cranial nerves palsy was suspected. An empiric treatment with intravenous administration of acyclovir for 10 days was carried out along with hydrocortisone for 10 days with gradual reduction of dosage, we also prescribed vasodilators and vitamins, and for management of skin lesions and prevention of secondary infection, we used Vaseline.

As a result of the treatment, the skin lesions disappeared first, then the patient regained a normal hearing.

After discharge facial palsy recovered with re-education. Hoarseness and dysphagia improved with the help of speech therapist. But full recovery was not obtained.

DISCUSSION

Herpes zoster, also known as shingles or zona, results from reactivation of latent varicella-zoster virus (VZV) infection within the ganglia of the spinal cord and cranial nerve usually in older and immunocompromised individual. It occurs years after having chicken pox. In the head and neck region, the most common presentation is Ramsay Hunt syndrome [1].

Ramsay Hunt syndrome is characterized by varicella zoster virus (VZV) infection affecting the geniculate ganglion of the facial nerve (VII). It typically presents with an erythematous vesicular rash in the external auditory canal or in the mouth associated with auricular pain and peripheral facial nerve paralysis. Vestibulocochlear nerve (VIII) is frequently co-involved during the course of Ramsay Hunt syndrome, which could be explained by the close proximity of the geniculate

ganglion to the vestibulocochlear nerve within the bony facial canal [2].

However multiple cranial nerves involvement has rarely been described in the literature [3]. Three different theories are proposed to explain multiple nerves involvement. The first theory is spreading of the virus through cerebrospinal fluid or by haematogenous way. The second theory is the presence of anastomoses between cervical nerves and branches of the facial nerve as an anatomical variation, leading to the spread of inflammation along these anastomoses. The third theory suggests a simultaneous activation of the virus in more than one ganglion [4].

VZV infection could cause a spectrum of symptoms; various combinations of cranial nerves can be involved [3,5,6], the skin and mucosal eruptions can be absent [7]. Therefore, clinicians should consider VZV infection in case of unilateral multiple cranial neuropathies regardless of whether vesicular skin or mucosal lesions are present or absent.

The diagnosis of zona is clinical in the vast majority of cases. But in the absence of vesicles, virological documentation of VZV infection seems essential; such as direct detection of virus DNA or antigens from vesicle liquid, body fluid (serum or cerebrospinal fluid) or tissue and serological tests for antibody can be performed which is considered positive if IgM antibodies against VZV are found, however, IgG values lack of diagnostic specificity but have some follow-up interest as that they reach their highest level 3 weeks after disease onset [8].

CT-scanner is usually normal but it may be performed if an acute cerebral infarction or a tumour were suspected. MRI occasionally showed an enhancement of the involved cranial nerves [3].

Treatment is based on the association acyclovir and corticosteroids, which must begin within 3 days of the onset of the disease for better recovery result [9]. Combined treatment has showed better results than monotherapy [10]. Acyclovir prevents further spread of the virus and corticosteroids reduce neural oedema [9]. However, new studies suggest that brivudin, famciclovir, and valacyclovir are expected to have better efficiency than acyclovir [11].

CONCLUSION

Herpes zoster is a rare medical condition necessitating early diagnosis that should be evoked in case of unilateral multiple cranial neuropathies regardless of whether skin or mucosal lesions exist or not. The combination therapy with antivirals and corticosteroids must be initiated as soon as possible in hope to obtain full recovery.

Conflicts of interest: Authors have declared that no competing interests exist.



REFERENCES

1. Hoshino C, Akane Y. Where is reactivation after a long latency? *BMJ Case Reports*. 2012; 10: 11-36.
2. Sweeney CJ, Gilden DH. Ramsay hunt syndrome. *J NeurolNeurosurg Psychiatry*. 2001;71 (2): 149–154.
3. Nisa L, Landis BN, Giger R, Leuchter I. Pharyngolaryngeal Involvement by Varicella-Zoster Virus. *J Voice*. 2013; 27 (5): 636-41.
4. Kayayurt K, Yavasi O, Bilir O, Ersunan G, Giakoup B. A Case of Ramsay Hunt Syndrome with Atypical Presentation. *Turk J Emerg Med*. 2014; 14 (3): 142-5.
5. Shinha T, Krishna P. Ramsay Hunt syndrome and zoster laryngitis with multiple cranial nerve involvement. *IDCases*. 2015; 2(2): 47-8.
6. Shihada R, Brodsky A, Luntz M. Laryngeal Mass with Multiple Cranial Neuropathies as a Presenting Sign for Varicella Zoster Infection. *Dysphagia*. 2010; 25(2): 153-5.
7. Gilden D, Cohrs RJ, Mahalingam R, Nagel MA. Neurological Disease Produced by Varicella Zoster Virus Reactivation Without Rash. *Curr Top MicrobiolImmunol*. 2010; 34: 243-53.
8. Watelet J-B, Evrard A-S, Lawson G, Bonte K, Remacle M, Van Cauwenberge P, et al. Herpes zoster laryngitis: case report and serological profile. *Eur Arch Otorhinolaryngol*. 2007; 264 (5): 505-7.
9. Murakami S, Hato N, Horiuchi J, Honda N, Gyo K, Yanagihara N. Treatment of ramsay hunt syndrome with acyclovir-prednisone: Significance of early diagnosis and treatment. *Ann Neurol*. 1997; 41(3): 353–357.
10. Kinishi M, Amatsu M, Mohri M, Saito M, Hasegawa T, Hasegawa S. Acyclovir improves recovery rate of facial nerve palsy in Ramsay Hunt syndrome. *AurisNasus Larynx*. 2001; 28(3): 223–226.
11. Dworkin RH, Johnson RW, Breuer J, Gnann JW, Levin MJ, Baconja M, et al. Recommendations for the management of herpes zoster. *Clin Infect Dis*. 2007; 44(Supp1): 1–26.