# Polineuropatia atípica causada pelo vírus varicela Zoster: Envolvimento dos nervos vestibulococlear, glossofaríngeo e vago

Atypical polyneuropathy caused by varicella Zoster virus: Involvement of vestibulocochlear, glossopharyngeal and vagus nerves

Mafalda Silva Ferreira • Joana Pires • Filipa Carvalho • Ana Margarida Amorim • Luís Silva

### RESUMO

O vírus varicela-zoster permanece latente no nosso corpo após uma infecção primária. O herpes zoster aparece após a sua reativação. A apresentação mais comum deste tipo de vírus, na região da cabeça e pescoço, é a síndrome de Ramsay Hunt (paralisia facial, herpes auricular e perda auditiva). Relatamos o caso de uma doente com uma manifestação atípica e rara de herpes zoster, que envolveu concomitantemente os nervos cranianos VIII, IX e X, preservando o VII.

Palavras-chave: Virus varicella zoster, Varicela, Herpes zoster, Paralisia da corda vocal.

Mafalda Silva Ferreira ENT Department, Coimbra University Hospital

Joana Pires ENT Department, Coimbra University Hospital

Filipa Carvalho ENT Department, Coimbra University Hospital

Ana Margarida Amorim Vestibular Unit, ENT Department, Coimbra University Hospital

### Luís Silva

ENT Department, Coimbra University Hospital

### Correspondência:

Mafalda Silva Ferreira ENT Department, Coimbra University Hospital Rua Professor Mota Pinto 3075, Coimbra, Portugal. E-mail: Mafalda.sf1990@gmail.com

Artigo recebido a 4 de Dezembro de 2018. Aceite para publicação a 18 de Março de 2019.

### ABSTRACT

Varicella-zoster virus stays latent in our body after a primary infection. Herpes zoster appears after its reactivation. The most common presentation of this type of virus, in head and neck region, is the Ramsay Hunt syndrome (facial palsy, otic herpes and hearing loss). We report a case of a patient with an atypical and rare herpes-zoster manifestation which envolved concomitantly the VIII, IX and X cranial nerves, preserving the VII. Keywords: Varicella zoster virus, Varicella, Herpes zoster, Vocal fold paralysis.

### INTRODUCTION

Varicella is a common pediatric disease, characterized by fever and a generalized pruritic vesicular rash. It results from a primary infection by varicella-zoster virus (VZV). VZV becomes latent in cells of the dorsal root ganglia after primary infection. Unlike its first presentation, herpes zoster reactivation affects a restrict area, manifesting itself as an extremely painful vesicular rash involving one or more adjacent dermatomes. The incidence of VZV is higher in the elderly and immunosuppressed individuals. This type of infection is an important cause of cranial neuropathy commonly affecting facial and vestibulocochlear nerves. Despite these frequent locations, affection of other cranial nerves is rarely seen. Since there are such uncommon manifestations, we present an atypical case of VZV infection affecting the vestibulocochlear, glossopharyngeal and vagus nerves.<sup>1,2,3</sup>

### **CASE REPORT**

A 68-year-old female patient was firstly observed in a peripheral hospital complaining of a sore throat, right ear otalgia, right fronto-parietal headache, dizziness and general malaise. As comorbidities she presented hypertension and dyslipidemia, medically controlled. On this first examination, vesicular eruptions were observed on the right ear, with a remaining normal ENT exam. She was medicated with oral brivudine 125mg once a day, analgesics and a multivitamin complex. As she was feeling worse, she returned to the hospital one week later, now complaning of otorrhea, fever, dysphonia, dysphagia and vertigo. A cerebral computed tomography scan (CT scan) was requested and merely revealed the presence of micronodular calcifications at the right parietal sulcus.

Thereafter she was transfered to a terciary hospital. Clinical observation revealed auricular (fig.1) and tympanic herpetic eruptions, otorrhea, paresis of the right soft palate (fig.2) and right vocal fold (fig.3) with a diminished gag reflex on the same side. No

## FIGURA 1

Otic herpes.



**FIGURA 2** Soft palate paresis.



**FIGURA 3** Vocal fold paresis.



spontaneous nystagmus was found, but a positive head shaking test with a left beating horizontal nystagmus and a right positive head impulse test were present. She was hospitalized for a period of 19 days. No facial paralysis was documented during hospitalization. She was submitted to a lumbar puncture and a gastroendoscopy which showed no alterations. A new head and neck CT just revealed filling of the right middle ear. Audiometry showed a mild sensorineural hearing loss on the right side and a mild presbycusis on the left. Videonystagmography (evaluation of occular mobility and optokinetic nystagmus, rotational chair test) denoted a left preponderance. Also during this period an head MRI was requested. The analytical and serological study confirmed the presence of an active infection by VZV.

During the admittance the treatment established consisted in 250 mg of oral acyclovir, multivitaminic complex (cyanocobalamin, pyridoxine, thiamine), 30 mg per day of deflazacort, 1 gr per day of ceftriaxone (5 days), gabapentin 100 mg three times per day and analgesy. The patient also attended speech and swallowing therapy sessions. She was discharged after a period of 19 days with a clinical improvement, mainting only a soft palate and vocal fold paresis on the ENT evaluation.

### DISCUSSION

We report an unusual case of an imunocompetent but elderly patient with herpes zoster oticus with multiple cranial nerves involvement.

Ramsay Hunt Syndrome classically comprises a painful vesicular rash on the external ear (herpes zoster oticus), associated with ipsilateral lower motor neuron facial nerve palsy, which appears some days before or after the rash. In this case the patient presented a painful vesicular rash on the right ear, right soft palate and right vocal fold paralysis (IX and X affection). Also, hearing loss and nystagmus were observed, indicating the VIII cranial nerve involvement as well. Surprisingly, facial asymmetry was not observed putting aside VII nerve impairment.

Some theories try to explain the viral spread and the implication of several cranial nerves. One theory postulates that there are neural anastomosis between the V, VII, IX, and X cranial nerves at the auricle and external auditory canal, therefore zoster infection can be disseminated between these pairs through this region. Other theory is vasculitis. Vascular anastomosis, from the infected artery, can disseminate the virus through the bloodstream, and it is known that vascular damage is common during herpes zoster infection, affecting both small and large arteries.<sup>4</sup> Thus, it is possible to explain the presence of hypoacusia, nystagmus, palatal and vocal fold paralysis with vesicular skin eruptions in the same patient.

In this case, the improvement of hearing loss was unsatisfactory with the applied corticotherapy. In literature, other cases report that hearing loss was less responsive to treatment comparatively to the other disorders. Vertigo resolved positively with medication and rehabilitation.<sup>5,6</sup>

The neck CT scan and laryngoscopy excluded other causes for soft palate and laryngeal paralysis. In this kind of nervous condition, we must be alert to the possibility of dysphagia often accompanied by aspiration. Speech and swallowing therapy sessions substantially improves the quality of life and prognosis of this kind of patients. Still, a preventive percutaneous endoscopic gastrostomy should be considered in some cases, even during a favorable evolution.

Since our patient complained about intense headaches, resistant to analgesia, we decided to do a lumbar puncture and a CT scan to rule out meningeal spread.

Lastly, we recognize that serological tests provides an important diagnosis confirmation and gives confidence to initiate an early antiviral and corticotherapy treatment, in order to maximize the recovery rate of the nerve function.<sup>7</sup>

### Protecção de pessoas e animais

Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

### Confidencialidade dos dados

Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

### Conflito de interesses

Os autores declaram não ter nenhum confito de interesses relativamente ao presente artigo.

### Fontes de financiamento

Não existiram fontes externas de financiamento para a realização deste artigo.

#### Referências bibliográficas:

1.Rasmussen ER, Mey K. Vocal cord paralysis associated with Ramsay Hunt syndrome: looking back 50 years, BMJ Case Rep. 2014;2014(feb06):

www.casereports.bmj.com/content/2014/bcr-2013-201038

2.Adachi M. A Case of Varicella Zoster Virus Polyneuropathy: Involvement of the Glossopharyngeal and Vagus Nerves Mimicking a Tumor. AJNR Am J Neuroradiol, 2008;29(9):1743-1745

3.Lee D, Yoon T, Lee J, Joo Y, et al. Herpes Zoster Laryngitis Accompanied by Ramsay Hunt Syndrome, J Craniofac Surg, 2013;24(5):e496-e498

4.Nishioka K, Fujishima K, Kobayashi H, Mizuno Y, et al. An extremely unusual presentation of varicella zoster viral infection of cranial nerves mimicking Garcin syndrome. Clin Neurol Neurosurg, 2006;108(8):772-774

5.Lee H, Yeh C, Hung S. Ramsay Hunt syndrome with vocal fold paralysis. Kaohsiung J Med Sci, 2014;30(5):264-265

6.Gómez-Torres A, Medinilla VA, Abrante JA, Esteban OF. Ramsay-Hunt Syndrome presenting laryngeal paralysis. Acta Otorrinolaringol Esp (English Edition), 2013;64(1):72-74

7.Fujiwara K, Furuta Y, Fukuda S. A Case of Associated Laryngeal Paralysis Caused by Varicella Zoster Virus without Eruption. Case Rep Med, 2014;2014:1-3