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Atypical Presentation of Ramsay Hunt Syndrome without Facial Palsy in an Immunocompetent Senegalese Adulte Patient

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Abstract

First described in 1907 by James Ramsay Hunt, Ramsay Hunt syndrome is a recurrence (reactivation) of varicella-zoster virus (VZV) affecting the geniculate ganglion, secondary to a decrease in cell-mediated immunity. The strict definition of Ramsay Hunt syndrome is peripheral facial nerve palsy accompanied by erythematous vesicular rash on the ear. We report a 57-year-old female immunocompetent patient complaining of otalgia, small vesicles on the Ramsey Hunt Zone. She does not complain fever, hearing loss, nausea, vomiting or dizziness. There was no peripheral facial nerve palsy, no reduction of taste sensation, no ataxia or nystagmus, Romberg sign was negative. Our patient targets two of the three criteria needed for the diagnosis of Ramsay Hunt syndrome. She began to take Acyclovir-Steroid (AS) therapy very early with good outcome. This suggests that prompt diagnosis and management improve outcome and prevent occurrence of nerve palsy in Ramsay Hunt syndrome.

Keywords

Ramsay Hunt Syndrome, Varicella Zoster Virus, Immunocompetent, Antiviral-Steroid Therapy

1. Introduction

Ramsay Hunt syndrome is a reactivation of the varicella-zoster virus (VZV) in the geniculate ganglion of the facial nerve [1]. It is characterized by clinical polymorphism, its self-limiting evolution, sometimes it's responsible for neurological permanent damages. Usually symptoms associate, peripheral facial palsy, and

vesicular eruptions on the Ramsay Hunt zone. The most common sign is peripheral facial palsy. Immunodepression increases the risk of occurrence. Prompt diagnosis and management improve outcome in Ramsay Hunt syndrome. We reported an atypical form of Ramsey Hunt Syndrome in an immunocompetent patient in whom facial palsy was not found.

2. Case Report

This was a 57-year-old female patient with known hypertension for 2 years treated with amlodipine 5 mg per day; she had been complaining of left otalgia, skin rash with preceding burning pain. However, she does not complain of fever, hearing loss, nausea, vomiting or dizziness. Physical examination found inflammatory swelling of the left side of the face, herpetiform vesicles in left ear, temporal, cheek, and mandibular regions (**Figure 1**); purulent otorrhea, an inflamed external auditory canal with a congestive tympanic membrane. Ophthalmological examination revealed no keratitis lesions. There was no reduction of taste sensation, no ataxia in walking or nystagmus, Romberg sign was negative.

There were no obvious abnormalities in renal function. Routine blood tests showed total neutrophil count of 12.06×10^9 /L, and inflammatory marker CRP of 312 mg/l. Syphilis, HIV, hepatitis B virus, and hepatitis C virus tests, were negative. Serum protein electrophoresis, fasting blood glucose (0.83 g/l) and HbA1c (6.5%) were all normal. She was treated with: Acyclovir-steroid and amoxicillin/ clavulanic. Otorrhea dried up in 3 days, vesicles had been absorbed and crusted after one week of treatment (**Figure 2**).



Figure 1. Unilateral aggregated vesicular rash at the beginning of the disease (2023.01.13).



Figure 2. Vesicular crusted at the end of the treatment (2023.01.20).

3. Discussion

Ramsay Hunt syndrome is the second most common cause of atraumatic peripheral facial palsy and involves approximately 5 cases per 100.000 people. Compared with Bell palsy, Ramsay Hunt syndrome generally has more severe paralysis at onset and patients are less likely to recover completely [1] [2]. It is caused by reactivation of VZV in the geniculate ganglion of the facial nerve. The most common clinical symptoms are a combination of peripheral facial palsy, a painful erythematous vesicular rash affecting the outer and inner ear, and otalgia [3].

In our patient, we didn't find the complete Ramsay Hunt syndrome. This can be explained by the fact that several nerves share the sensitive innervation of Ramsay Hunt's area (IX and X at the same time as Wrisberg's intermediary) [4]. Thus, damage to the auricular zone of Ramsay Hunt may involve only one or other of the nerves responsible for innervation. This is why clinicians should suspect Ramsay Hunt Syndrome when there is only one sign of this classic triad, to avoid any delay in diagnosis.

When faced with a rash, the differential diagnosis must be made with impetigo, which spreads more rapidly and extensively, herpes, and diffuse externa otitis, either simple or associated with otitis media [5]. Moreover, auditory, and vestibular involvement may occur without facial paralysis, with or without an eruptive syndrome [6]. This atypical form without peripheral facial nerve palsy occurred in our immunocompetent patient. It's rare and may require a virological and serological diagnosis [6] [7]. Our patient targets two of the three criteria needed for the diagnosis of Ramsay Hunt Syndrome, so we didn't use biology to confirm the diagnosis, because the diagnosis is fundamentally clinical [8] and

the lesions were without doubts vesicles.

Our patient was immunocompetent, that is why, in our opinion, he promptly returned to his normal life and functionality in only four days of antiviral therapy. In fact, age greater than 60 years, diabetes mellitus, essential hypertension and associated vertigo were identified as prognostic factors for worse outcome and decreased chance of recovery [9], hypertension is the one condition found in our patient. But most important, he begun to take acyclovir-steroid (prednisone) therapy (AS) very early, we think that it is the reason of the absence of neural facial palsy as suggested by several studies in which there is evidence that in normal patients without immunosuppression, early administration of acyclovir-prednisone would improve the rate of healing of the skin lesions, diminish the duration of pain of herpes zoster, furthermore was proved to prevent nerve degeneration by nerve excitability test [10] [11] [12]. Early administration of acyclovir-prednisone treatment within the first 3 days of the onset of the disease leads to complete resolution of symptoms [2]. We therefore initiated treatment within 24 hours of the onset of the disease and we added an antibiotic to treat purulent otorrhea, moreover, instillation of antibiotic-cortisone ear solutions is recommended in cases of otitis externa [5].

4. Conclusion

Ramsay Hunt syndrome is characterized by the occurrence of vesicular rash on the ear and peripheral facial nerve palsy. However, in relation to anatomical arrangements of the nerves that concomitantly innervate the Ramsay Hunt area, various clinical features may occur, such as the absence of peripheral nerve palsy observed in our patient. Nevertheless, there is a real risk of secondary progression to peripheral nerve palsy or post-herpetic neuralgia. Hence the importance of early diagnosis and antiviral treatment is to avoid this possibility. Prompt diagnosis and management improve outcome in Ramsay Hunt syndrome.

Consent

The patient has signed an informed consent form, which is disponible.

Authors' Contributions

DT and TAI wrote the manuscript with input of KDM and NAL. JZY collected and prepared the figures. MS drafted and approved the final version to be published.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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