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(CASE REPORT)

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A case report of Ramsay Hunt Syndrome involving the cervical dermatomes in Nigeria

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Abstract

A rare neurological disorder, Ramsay Hunt Syndrome is caused by reactivation of varicella-zoster virus in the geniculate ganglion. The syndrome typically comprises a triad of unilateral facial palsy, vesicular rash around the ear canal or mouth and otalgia. However, atypical cases occur posing diagnostic challenges and risk of poorer outcomes. The purpose of this report is to highlight an atypical presentation of this syndrome where published cases are limited. We describe a 56-year-old Nigerian woman who presented with incomplete triad of Ramsay Hunt Syndrome characterized by unusual cervical dermatomal distribution of the zoster rash, facial palsy and absence of otalgia. Despite initial missed diagnosis elsewhere, timely administration of Acyclovir and Prednisolone resulted in marked clinical improvement. The atypical presentation highlights the importance of considering Ramsay Hunt Syndrome in patients with facial palsy and a rash anywhere in the head and neck even if the classic triad is incomplete.

Keywords: Cervical Dermatomal Zoster Rash; Facial Palsy; Nigeria; Otalgia; Ramsay Hunt Syndrome

1. Introduction

Ramsay Hunt Syndrome (RHS), a rare neurological disorder with an incidence of 5 per 100,000 population/year. It was first described in 1907 and classically presents as a triad of facial paralysis, dermatomal vesicles around the auricle or mouth, and otalgia. [1, 2] The syndrome follows the reactivation of latent varicella zoster virus (VZV) infection in the geniculate ganglion.

Atypical cases of RHS are rare and could lead to missed diagnosis and complications.[3] This case report of atypical RHS with cervical dermatomal (C2/C3) rash and absence of otalgia aims to raise awareness of atypical presentations of RHS.

2. Case Report

A 56-year-old woman presented to a tertiary hospital in Nigeria with painful rash at the nape of her neck of two days duration. Pain was self-rated 8 on the visual analogue scale (VAS).[4] She denied auricular symptoms. The patient had chickenpox during childhood. She had hypertension, but not diabetes, HIV infection, facial trauma or burns. Her medication history consisted of the antihypertensive, Amlodipine and Ibuprofen for analgesia from another facility where the diagnosis was missed before the patient reported to our facility because of severe, unremitting pain on the left side of her neck without otalgia.

Her physical examination revealed a fully conscious and afebrile woman with left peripheral facial palsy, Grade III on House-Brackmann facial nerve grading system in Table 1.[5]

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Table 1 House-Brackmann facial nerve grading system.[5]

Grade of Facial Nerve Function	Characteristic Features
Grade I. Normal	Normal facial function in all areas.
Grade II. Slight Dysfunction	Gross: slight weakness noticeable on close inspection; may have very slight synkinesis; At rest: normal symmetry and tone; Motion: Forehead - moderate to good function; Eyes - complete closure with minimum effort; Mouth - slight asymmetry.
Grade III - Moderate Dysfunction	Gross: obvious but not disfiguring difference between two sides; noticeable but not severe synkinesis, contracture, and/or hemi-facial spasm; At rest: normal symmetry and tone; Motion: Forehead - slight to moderate movement; Eyes - complete closure with effort; Mouth - slightly weak with maximum effort.
Grade IV - Moderate Severe Dysfunction	Gross: obvious weakness and/or disfiguring asymmetry; At rest: normal symmetry and tone; Motion: Forehead - none; Eyes - incomplete closure; Mouth - asymmetric with maximum effort.
Grade V - Severe Dysfunction	Gross: only barely perceptible motion; At rest: asymmetry; Motion: Forehead - none; Eyes - incomplete closure; Mouth - slight movement;
Grade VI - Total Paralysis	No movement.

She had ipsilateral vesicular and crusting lesions at the left side of C2/C3 dermatomes of the head and neck, and a few at the helix of the left pinna (Fig. 1).

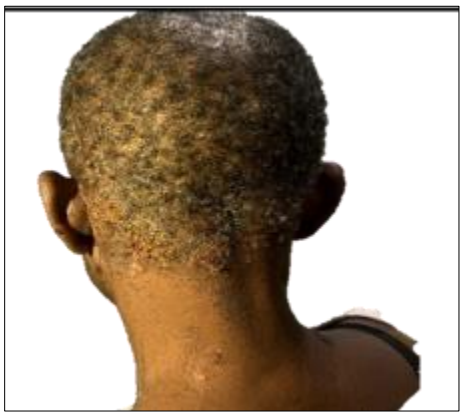

Figure 1 Zoster vesicles and crusts on the left ear and at the back of the head and neck of the patient

Photo credit: Corresponding author with patient's informed consent. June 2024.

No lesions were observed in her auditory canal or oral cavity.

The power of her limbs was normal and so was audiometry. Her blood pressure (BP) on presentation was 160/100 mmHg. Random blood glucose was 85 mg/dL and she was seronegative for human immunodeficiency virus (HIV). A clinical diagnosis of Ramsay Hunt Syndrome was made. Diagnostic tests for VZV were not available.

She was promptly treated on outpatient basis with oral Acyclovir 800 mg 5 times daily for 5 days and Prednisolone tablets at 60 mg daily for ten days. Tramadol and Carbamazepine were prescribed for pain relief. Her antihypertensive therapy was optimized. After two weeks, she had significant pain reduction from 8 to 3 on the VAS and facial palsy improved to Grade I on House-Brackmann grading system. The rash had also healed completely. The BP improved to 130/80 mmHg. Complications like postherpetic neuralgia and synkinesis were not discernible. Her clinical response was remarkable.

3. Discussion

Atypical presentation of RHS may lead missed or delayed diagnosis as in the present case, without the classic triad. The patient had no otalgia and the zoster lesions were located at the C2, C3 dermatomes suggesting involvement of the superior cervical ganglion. Cervical dermatomal involvement in RHS is rare, though previously reported in literature in a Caucasian woman. [1, 6] In RHS, VZV may infect several cell types in central and peripheral nervous system, which may explain the presentation in this case.[7]

A differential diagnosis for RHS is Bell's palsy, which however, does not present with zoster rash. The patient had hypertension, a leading risk factor for stroke, but stroke was clinically excluded because the facial palsy was peripheral and no limb paralysis. However, stroke-like syndromes may accompany RHS. [1, 7, 8]

Delayed treatment can lead to complications such as postherpetic neuralgia, synkinesis, hearing loss, exposure keratitis, and permanent hemifacial paralysis. Three of four patients are expected to recover fully from RHS, if offered timely treatment within 72 hours of onset, with a combination of antiviral drugs (Acyclovir, Famciclovir, Valacyclovir) and steroids (Prednisolone).[9] Contrastingly, only 30% of patients recover completely, if treatment is initiated after seven days of symptom onset.[9] Monotherapy with either agent is less effective than the combination. The effectiveness of nucleoside analogue antiviral agents like Acyclovir lies in their ability to block viral DNA. The anti-inflammatory action of steroids is most effective early when inflammation is most intense.

Varicella-associated complications such as RHS are higher among the unvaccinated and in low resource countries. This calls for improved access to effective recombinant zoster vaccines (RZV) in resource-limited settings like Nigeria to reduce VZV burden and prevent complications like RHS.[10]

4. Conclusion

In conclusion, this case report highlights a rare and atypical case of RHS and the importance of heightened awareness and timely management to enhance recovery and to prevent complications. Public health intervention with routine VZV vaccination is recommended to reduce the burden VZV infection and to prevent the disabling sequelae of RHS, which negatively impact quality of life. The report adds to the limited literature on RHS, underscoring the need for further research to better understand the epidemiology and clinical spectrum of Ramsay Hunt Syndrome.

Compliance with ethical standards

Acknowledgments

The authors wish to acknowledge the patient who voluntarily consented to have her case reported and photos taken for this case report.

Disclosure of conflict of interest

The authors declare that they have no conflicts of interest regarding this case report.

Statement of ethical approval

The authors sought and obtained ethical approval from the Igbinedion University Teaching Hospital Research Ethics Committee.

Statement of informed consent

The patient voluntarily provided informed consent for the publication of her case and accompanying photo.

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