

Case Report

Ramsay Hunt syndrome presenting as “a window” to HIV diagnosis in 48 year old African female: A case report

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ABSTRACT

Background: Ramsay Hunt syndrome in adults is reported in a number of case reports as an uncommon presentation of Varicella - zoster Virus infection. Ramsay Hunt syndrome occurs when there is reactivation of the latent Varicella - zoster virus present in the ganglia due to previous varicella infection. Following a breakdown of the immune system, latent Varicella - zoster infection can be activated. Human Immunodeficiency Virus infection (HIV), causes immunosuppression and hence reactivation of Varicella - zoster. The diagnosis of Ramsay Hunt syndrome, gives a window of opportunity to investigate for HIV. Here we report our first case of Ramsay Hunt syndrome as a presenting feature of HIV disease in an apparently healthy looking 48 year old black African female who was unaware of her HIV status at the time of presentation.

This case is reported in order to highlight the need to use Ramsay Hunt syndrome as a window of opportunity to screen for underlying HIV.

Case presentation: A 48 year old black African female who was unaware of her HIV status, presented to our outpatients department with a two day history of painful right ear pain, vesicles on the right pinna and inability to completely close her right eye, her mouth was deviated to the left, with a wrinkle free frown. She denied any alteration in taste sensation and hyperacusis. She had a

grade IV House-Brackmann facial paralysis. A diagnosis of Ramsay Hunt syndrome was made and an HIV test was offered, the test was positive for HIV. A subsequent Highly Active Antiretroviral Therapy (HAART) work up, revealed Chronic Kidney Disease stage 3B (CKD 3B) and a cluster of differentiation 4(CD4) count of 217 cells/ μ l. She was started on antiretroviral therapy 2 weeks after completing a 10 day regimen of acyclovir. In addition to her drug treatment she received ocular care and physiotherapy. Four months on her facial nerve function has improved modestly.

Conclusion: An HIV test should be an indispensable part of the Ramsay Hunt syndrome workup, as its diagnosis gives a window of opportunity to diagnose HIV and offer life- saving HAART.

INTRODUCTION

Ramsay Hunt syndrome occurs due to Varicella- zoster virus affecting the geniculate ganglion of the seventh cranial nerve that leads to a lower motor lesion of the facial nerve. It is characterized by a Bell's phenomenon, ear pain, and vesicles in the ipsilateral auricle and external auditory meatus.^{1, 2} Facial muscle paralysis can be assessed using the House - Brackmann staging.³ Varicella - zoster infection can be a feature of Acquired Immunodeficiency Syndrome (AIDS) stage 2.⁴ There are several case reports of Ramsay Hunt syndrome as an initial presenting feature of undiagnosed HIV. We report our first case of Ramsay Hunt syndrome in HIV at Livingstone Central Hospital, despite the high national HIV prevalence of 13%.⁵

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CASE PRESENTATION

A 48 year old black African female presented to our outpatients department with a 2 day history of painful right ear pain, vesicles on the right pinna and inability to completely close her right eye, her mouth was deviated to the left, with a wrinkle free frown. She denied any alteration in taste sensation and hyperacusis. She had a grade IV House-Brackmann facial paralysis. The rest of her clinical evaluation was otherwise unremarkable.

Figure 1. Time course: Ramsay Hunt syndrome over four months.

Panel A



Day 1: Right wrinkle free frown.

Panel B



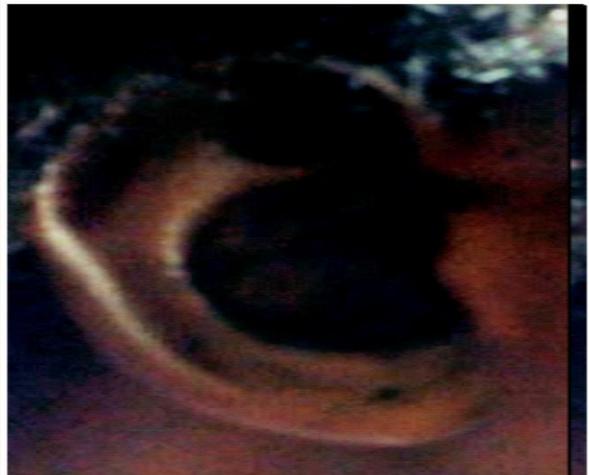
Day 1: Herpetic vesicles on right pinna

Panel C



Four months later: Unable to completely close the right eye

Panel D



Four months later: Healed herpetic vesicles.

She appeared well nourished with no obvious pointers to immunosuppression as she had no history of exposure to chemotherapy, prolonged corticosteroid use nor diabetes mellitus. Notably, she was unaware of her HIV status. Rapid determine and Unigold HIV tests came out positive. For the Ramsay Hunt syndrome, she received a 10 (ten) day course of acyclovir 800mg five times daily, physiotherapy, artificial tears and eye patch. Prednisolone was omitted in view of her advanced immunosuppression and controversies associated with steroid use in Ramsay Hunt syndrome.^{6, 7} Within a fortnight, we initiated her on HAART with abacavir, lamivudine, efavirenz as her antiretroviral regime as work-up revealed advanced HIV disease with a baseline CD4 count of 217 cells/ μ l, CKD stage 3B (serum creatinine 122 μ mol/l, MDRD (Modification of Diet in Renal Disease Study) eGFR

(estimated glomerular filtration rate) of 43ml/min/1.73m², with bilateral small kidneys of 8.7cm x 10cm). She remains under our antiretroviral therapy outpatient clinic follow up and she shows a modest response as four months on, her House - Brackmann stage IV facial paralysis has improved to stage III. The auricular vesicles healed completely without scarring (Figure 1 and table 1). Her kidney function has improved from CKD stage 3B to CKD stage 2, MDRD eGFR of 68 mls/min/1,73m².

Table1: Timeline: Ramsay Hunt syndrome in HIV /AIDS clinical presentation and management over 4 months

Step	Time of presentation at the medical outpatient clinic	Clinical finding
A	Day1: First time presentation. <i>(acyclovir, paracetamol & codeine phosphate),ocular care and physiotherapy prescribed,HIV test positive and HAART workup initiated, with subsequent ARV initiation at 2 weeks visit-abacavir,lamivudine and efavirenz)</i>	Facial nerve paralysis: Lower Motor Neuron lesion type. She had a right wrinkle free frown. (House- Brackmann stage IV paralysis)
B	Day1: First time presentation. <i>(acyclovir, paracetamol & codeine phosphate),ocular care and physiotherapy prescribed, HIV test positive and HAART workup initiated, with subsequent ARV initiation at 2 weeks :abacavir,lamivudine and efavirenz)</i>	She had a vesicular rash on the right pinna.
C	Four months later: <i>House-Brackmann staging used to assess facial nerve function recovery</i>	Frown wrinkle had returned but she was unable to completely close her right eye. (House- Brackmann stage III paralysis.)
D	Four months later: <i>House-Brackmann staging used to assess facial nerve function recovery</i>	The vesicles on the right pinna were completely healed without scarring.

described as a clinical predictor of Human Immunodeficiency Virus.⁸ The risk of developing herpes zoster depends on the decline of cell-mediated immunity therefore other causes of decreased cell mediated immunity like use of immunosuppressants ,old age or diabetes mellitus should be actively looked for in addition to HIV.

In our patient, Ramsay Hunt syndrome occurred at a CD4 count of 217 cells/μl; however Ramsay Hunt syndrome does not appear to have a set CD4 count at which it occurs. In one review, 10 cases of Bell's phenomenon in acute HIV type 1

infection occurred at variable CD4 counts.

Treatment with oral acyclovir was initiated in our patient because she presented within 48 hours after the onset of the auricular rash. Initiation of acyclovir within 48 hours of onset of the rash is needed for optimal efficacy as it shortens the time to complete resolution of zoster associated pain.¹⁰ In addition to pharmaceutical agents [Acyclovir with or without prednisolone and HAART], physiotherapy and eye care are an essential component to treatment and can lead to recovery of facial muscle paralysis and prevent development of exposure keratitis respectively.

DISCUSSION

In our environment, Ramsay Hunt syndrome is an uncommon presentation of herpes zoster, an HIV/AIDS Stage 2 illness. In settings of high HIV prevalence clinicians should screen for HIV in patients presenting with Ramsay Hunt syndrome as herpes zoster has been

CONCLUSION

Herpes zoster is an AIDS defining illness. Despite herpes zoster being an AIDS defining illness it is uncommon for herpes zoster to present as Ramsay Hunt syndrome in HIV. The occurrence of a Bell's phenomenon accompanied by auricular vesicular rash in an apparently

healthy looking adult should prompt a clinician to look for underlying immunosuppression. In high HIV prevalence environments an HIV test should be an indispensable part of the Ramsay Hunt syndrome workup as it gives an opportunity to initiate HAART if one is HIV positive and eligible. Physiotherapy and ocular care are as equally important as acyclovir with or without prednisolone in the treatment of Ramsay Hunt syndrome.

ABBREVIATIONS

HIV: Human Immunodeficiency Virus, HAART: Highly Active Antiretroviral Therapy, CKD: Chronic Kidney Disease, CD4: Cluster of Differentiation 4, MDRD: Modification of Diet in Renal Disease Study, AIDS: Acquired Immunodeficiency Disease Syndrome.

DECLARATIONS

Ethics approval and consent to participate

Not applicable to this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available with the Editor –in-Chief.

Availability of data and material

The authors are agreeable to make available the raw data and edited images described in our manuscript freely available.

Competing interests

The authors declare that they have no competing interests.

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Authors` contributions

All the three authors attended to the patient, contributed in writing, revised and approved the final manuscript.

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