Case study

Ramsay Hunt Syndrome with Cranial polyneuropathy and aseptic meningoencephalitis: *A case report*

Abstract

Introduction

Ramsay Hunt Syndrome is characterized by paroxysmal ear pain, vesicular rash around the auricle and ear canal with ipsilateral peripheral fascial nerve palsy. It is due to reactivation of the Varicella Zoster Virus (VZV) in the fascial nerve ganglion. It is considered the second most common cause of peripheral fascial nerve palsy. Involvement of other cranial nerves is rare. Dissemination of infection can also lead to meningoencephalitis which too is rare.

Case Presentation

A 14-year-old immunocompetent boy presented with fever associated with vertigo, fascial asymmetry and difficulty in swallowing. Careful history revealed that he had a rash on his left ear with ear pain one week prior to this. He had a few crusted lesions in his left ear pinna and ear canal on examination. Cranial nerve examination revealed a left sided fascial and palatal -palsy. Cerebrospinal fluid (CSF) analysis was suggestive of viral meningoencephalitis. He was treated for Ramsay Hunt Syndrome with cranial polyneuropathy and meningoencephalitis with Acyclovir and steroids. He made a successful recovery.

Conclusion

This case highlights the importance of clinicians recognizing Ramsay Hunt syndrome amongst patients presenting with multiple cranial nerve involvement and amongst those with meningoencephalitis. It is also a reminder that careful history and meticulous examination can lead to prompt recognition of a reversible cause of cranial polyneuropathy.

Keywords: Ramsay Hunt Syndrome, Cranial Nerves, Acyclovir, Varicella Zoster, meningoencephalitis

Introduction

Ramsay Hunt syndrome (RHS), first described by J. Ramsay Hunt in 1907, refers to the association of unilateral peripheral facial nerve palsy and reactivation of Varicella Zoster Virus (VZV) along the sensory nerves innervating the ear [1]. RHS has an incidence of around 5 per 100,000 and known to account for around 12% of fascial Nerve Palsies [2] Although the fascial neve is most commonly implicated other cranial nerve involvement too is possible, which include VII, VIII, IX, X, V and III/XI in descending order of involvement. The disease occurs due to reactivation of the VZV in the geniculate ganglion of the fascial nerve. It can be confirmed by demonstration of VZV DNA in the geniculate ganglion [3] Hunt explained the mechanism of multiple cranial nerve palsy as an involvement of adjacent ganglia by contiguous anatomical

contact from the original source of inflammation in a single ganglion [4] Another theory was that the spread of the virus through a common blood supply to cranial nerves. Other associations of RHS are Horner's syndrome, herpes zoster uveitis, meningoencephalitis, and the syndrome of inappropriate secretion of antidiuretic hormone. [5][6][7]

This case is unique as the child had both cranial polyneuropathy as well as meningoencephalitis both being uncommon associations of RHS. Such complications usually occur amongst immunocompromised patients. But this case report highlights the need to consider the possibility of RHS amongst even immunocompetent patients presenting with multiple cranial nerve involvement and meningoencephalitis.

Case Report

A 14-year-old previously healthy boy was admitted with a complaint of vertigo and fever for one week. One week prior to fever he also complained of a painful rash on his left ear which had resolved spontaneously. He described the rash to be clusters of small drops like lesions over his left pinna and ear canal. He also complained of pain in his left ear and tinnitus. He had fever during that week which was low grade in nature not associated with chills and rigors. He also complained of dysphagia and nasal regurgitation for one day and imbalance. He denied any weakness of his limbs. He had no headache, photophobia, phonophobia or neck stiffness. He had no fits or bladder/bowel incontinence. He had chicken pox when he was 7 years old which was uncomplicated. He had no significant past medical, surgical or allergic history. He is a school going child with average school performance. He denies smoking, alcohol or substance abuse.

On examination he was thin built and not in any respiratory distress. He had a left sided lower motor neuron type fascial nerve palsy (Figure 1) and a left sided palatal palsy (Figure 2). Eighth nerve examination revealed normal hearing with no sensorineural or conductive hearing losses. He had an ataxic gait with swaying to the left. Romberg's test was positive. Rest of the cranial nerve examination was normal. He had no other cerebellar signs and upper and lower limb examination too was normal. He had no neck stiffness and Kernig's sign was negative. Ear examination also revealed crusted lesions of the pinna and external auditory canal suggestive of a recent Herpes Zoster infection (Figure 3). There were few vesicles on the left side of palate as well. Rest of the systemic examination was normal.

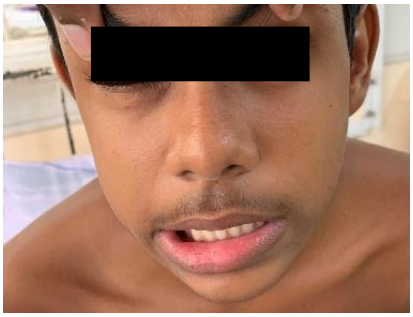

Figure 1

Morphological view

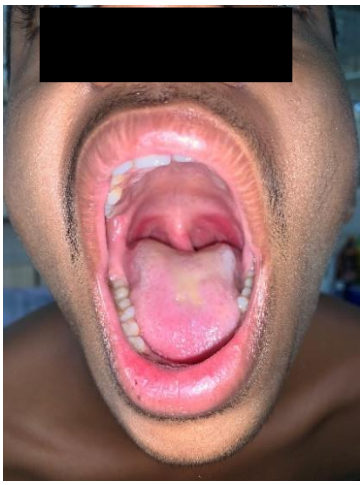

Figure 2

left sided palatal palsy

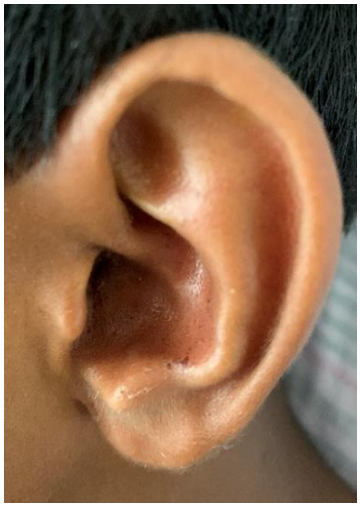

Figure 3

Herpes Zoster infection

Routine blood investigations were normal. Pure Tone Audiometry showed no hearing loss bilaterally.

Cerebrospinal Fluid (CSF) analysis results are shown in Table 1.

MRI Brain was normal with no evidence of tumor, infection or demyelinating disease.

A clinical diagnosis of Ramsay -Hunt Syndrome with involvement of seventh, eighth and tenth cranial nerves and VZV meningoencephalitis was made. He was started on IV Acyclovir 10 mg/kg three times a day for 14 days along with prednisolone 1mg/kg/day for ten days which was then tapered off.

Fascial weakness on House Brackmann scale was grade 3 (Moderate) at presentation and grade 2 (mild) on discharge. He was reviewed after one month and the fascial and palatal weakness had completely resolved. He no longer had vertigo or other features of vestibular component of eighth nerve involvement.

Protein	73 mg/dl
Cell Count	86/mm3
Neutrophils	1/mm3
Lymphocytes	85/mm3
CSF Sugar	68 mg/dl
Random Blood Sugar	96 mg/dl
Varicella Zoster Virus PCR	Positive
Herpes Simplex Virus 1 and 2 PCR	Negative
CytomegaloVirus PCR	Negative
Tuberculosis Gene Xpert	Negative
CSF culture	Negative

Test results

Table 1

Discussion

Reactivation of the VZV in the geniculate ganglion causing lower motor neuron fascial paly is a well described phenomenon and a common presentation in clinical practice. Involving of other cranial nerves although less common has been reported in literature. This patient had involvement of the ipsilateral tenth cranial nerve suggested by the left sided palatal palsy. Involvement of the eight cranial nerve too was a likely cause for his vertigo and positive Romberg's test. Normal hearing suggested involvement of only the vestibular component of the eight cranial nerve. It has been reported that the

incidence of vestibular nerve involvement is three- to four-fold higher than that of cochlear nerve involvement when cranial nerve VIII is associated with RHS [8]. Patient also had CSF fluid analysis suggestive of meningoencephalitis with CSF being positive for VZV DNA. The patient's clinical presentation was possibly due to that spreading of reactivated VZV causing local meningoencephalitis and multiple cranial nerve involvement. This is a rare combination of symptoms with a few cases reported previously [9] Further literature review showed that such extensive spread of the virus was rarely seen in immunocompetent patients such as this patient [10]

Due to the widespread involvement of unilateral cranial nerves, the possibility of malignant infiltration or sarcoidosis was considered although unusual at his age. Screening for these entities were negative.

The beneficial evidence of antiviral therapy in RHS still remains controversial. This patient was started on early antiviral treatment as his CSF analysis was suggestive of a viral meningoencephalitis, which is associated with high mortality and morbidity with delays in treatment. Treatment with acyclovir reduces the duration of symptoms [11]

MRI and other neuroimaging being normal in such patients have been reported before [12] This highlights the fact the Ramsay Hunt Syndrome with cranial polyneuropathies should be diagnosed clinically.

Conclusion

Peripheral fascial nerve palsy is a relatively common presentation in clinical practice and physicians should always consider the possibility of Ramsay Hunt syndrome amongst such patients. Involvement of multiple cranial nerves should not diverge the diagnosis from RHS but should rather be supportive of it. At the same time proper neurological assessment of other cranial nerves in patients with obvious fascial nerve palsies remain crucial. This case report also highlights the possibility of coexisting meningoencephalitis in such patients and the need for performing CSF analysis in patients with RHS. This will allow early initiation of anti-viral drugs that will significantly improve the outcome of such patients.

Patient Perspective

The patient declared that he understood that it was an infection that had caused all his symptoms and was compliant with the medication. He was satisfied with the treatment and happy that he made a good recovery

Informed Consent

Informed written consent was obtained from the patient's father to publish details regarding the patient's condition including photographic evidence.

References

[1]Sweeney CJ, Gilden DH "Ramsay Hunt syndrome" *J Neurol Neurosurg Psychiatry. 2001;71:149–154*

[2] Peitersen E. "Bell's palsy: the spontaneous course of 2,500 peripheral facial nerve palsies of different etiologies." *Acta Otolaryngol Suppl. 2002:4–30.*

[3] Van de Steene V, Kuhweide R, Vlaminck S, Casselman J. "Varicella zoster virus: beyond facial paralysis." *Acta Otorhinolaryngol Belg. 2004;58:61–66.*

[4] Hunt JR. "The symptom-complex of the acute posterior poliomyelitis of the geniculate, auditory, glossopharyngeal and pneumogastric ganglia." *Arch Intern Med. 1910;5:631–675*

[5] Aviel A, Marshak G. "Ramsay Hunt syndrome: a cranial polyneuropathy." *Am J Otolaryngol. 1982;3:61–66.*

[6]. Kageyama Y, Nakamura M, Sato A, Sato M, Nakayama S, Komatsuzaki O, et al. "Syndrome of inappropriate secretion of antidiuretic hormone (SIADH) associated with Ramsay Hunt syndrome: report of a case and review of the literature." *Jpn J Med. 1989;28:219–222.*

[7] Bhattacharyya PC, Kakati S. "Ramsay Hunt syndrome with aseptic meningitis." *J Assoc Physicians India. 1993;41:113–114*

[8]. Turner JE, Geunes PM, Schuman NJ. "Cranial polyneuropathy--Ramsay Hunt's syndrome: case report and discussion" *Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1997;83:354–357*

[9] A case of Ramsay Hunt syndrome associated with local meningitis, multiple cranial neuropathy, and the second cervical nerve involvement].

[10] Habib AA, Gilden D, Schmid DS, Safdieh JE. "Varicella zoster virus meningitis with hypoglycorrhachia in the absence of rash in an immunocompetent woman." *J Neurovirol. 2009;15(2):206–208*

[11] Gnann JW, Jr, Whitley RJ." Clinical practice. Herpes zoster." *N Engl J Med. 2002;347(5):340–346*

[12] Dyachenko PA, Dyachenko AG. "A case of MRI negative Herpes Virus Encephalitis presented by Ramsay Hunt Syndrome" *Wiad Lek. 2020;73(11):2555-2556.*