Ramsey Hunt Syndrome as a cause of Unilateral Facial Palsy in a Vaccinated Child

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ABSTRACT

Objective To discuss the differential diagnosis of Unilateral Facial Palsy in the Pediatric Population, and the importance of considering Varicella Zoster Virus (VZV) as an etiologic agent.

Design Case Report and Review of the Literature

Setting Tertiary Care Pediatric Referral Center

Conclusion Varicella Zoster Virus is a well-known cause of human viral infections including chicken pox and shingles. Despite effective pediatric vaccination, viral reactivation at the geniculate ganglion can result in Ramsey Hunt Syndrome type II and associated unilateral facial palsy.

CASE REVIEW

• 15 year old male presented with: a 3 day history of complete right sided facial palsy and a five day history severe right sided otalgia. Additionally, he reported mild right sided hearing loss and dysgeusia.

• PMH: no previous episodes of varicella or facial palsy reported. Vaccination schedule was up to date with varicella vaccinations in 1999 and 2009.

• External Ear: Dry, crusted lesion within the right conchal bowl. Right pinna was noted to have desquamation, edema and erythema, with extreme tenderness to palpation. External auditory canal was stenotic and erythematous, impeding clear examination of the tympanic membrane.

• CN VII: Right facial nerve palsy; VI/VI on the House-Brackmann scale.

• Serologic testing: VZV Ab IgM value within normal limits and a reactive VZV Ab IgG. Herpes simplex virus 1 and 2 IgG and IgM Ab were not detected and Lyme titers were negative.

• Diagnosis: Ramsay Hunt Syndrome type II, or Herpes Zoster Oticus

• Treatment: High dose IV methylprednisolone and IV acyclovir, strict eye care regimen. Patient was managed to complete recovery.

IMAGING

Figure 1: A: Pre-contrast T1 weighted MRI showing internal auditory canal and lateral semicircular canal. Arrow points to terminal portion of internal auditory canal. B: Post-gadolinium contrast T1 weighted MRI shows enhancement of facial nerve extending from lateral internal auditory canal to geniculate ganglion.

DISCUSSION

• Facial Palsy is a rare diagnosis in the Pediatric Population, occurring at a rate of 8.6 to 21.1 per 100,000 pediatric patients.1,2,3

• Ramsay Hunt Syndrome is a peripheral facial neuropathy accompanied by an erythematous vesicular rash of the ear, and is postulated to be the cause of 16.7% of pediatric facial palsy. It is caused by reactivation of varicella zoster virus (VZV) at the geniculate ganglion.4

• Zoster in children immunized with varicella vaccine is not always caused by the varicella Oka vaccine strain, and causative viruses revealed by PCR analysis to be wild strains of VZV. In this manner, children may still develop facial palsy secondary to VZV despite effective vaccination.5 Serology analysis is essential for the diagnosis of peripheral facial palsy due to VZV. To detect VZV, a twofold or greater rise in anti-VZV IgG titer is more useful than detection of anti-VZV IgM antibody.6

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