Unilateral Facial Swelling Caused by Ramsay Hunt Syndrome Resembles Odontogenic Infection

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SOMMAIRE

La cellulite faciale et l'œdème de la tête et du cou sont des signes d'infection odontogène inquiétants, qui peuvent mettre la vie du patient en danger. Bien que la plupart des infections de la tête et du cou soient causées par des pathogènes bactériens, les dentistes doivent également envisager la possibilité de maladies virales ou fongiques et les présentations qui y sont associées, au moment de traiter ces infections. Le présent rapport documente un cas d'infection virale qui ressemblait au départ à une infection odontogène bactérienne. Il vise à familiariser les dentistes avec la maladie de Ramsay Hunt et à les sensibiliser à l'importance de reconnaître rapidement cette maladie et d'instaurer un traitement définitif précoce.

Mots clés MeSH : cranial nerves/virology; diagnosis, differential; focal infection, dental; herpes zoster oticus/diagnosis

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Ramsay Hunt syndrome (RHS) is caused by the reactivation of a previous *Varicella zoster* virus (VZV) infection. RHS is a potentially serious viral infection that accounts for approximately 12% of all facial nerve palsies.[1,2] VZV is also the cause of "shingles," which frequently presents with a classic painful dermatomal distribution of vesicles and crusted skin ulcerations. Following initial viral exposure and inoculation, VZV may be found to reside in spinal nerve ganglia or in the cranial ganglia of the fifth, seventh or eighth cranial nerve.[3] The involvement of these cranial nerves may cause more severe complications than shingles alone: for example, unilateral facial paralysis or Bell's palsy.[4] In addition to the alarming facial palsy, RHS may also be characterized by severe otalgia, sensorineural hearing loss, vertigo, painful skin vesicles and aguesia in the ipsilateral anterior tongue.[5] RHS is rare in children, affects both sexes equally and is not contagious in individuals previously exposed to VZV.[6] Incidence and clinical severity increase when host immunity is compromised.[7]

The diagnosis of RHS is made primarily when clinical findings of a unilateral rash or vesicles in one facial dermatome exist in conjunction with unilateral facial palsy and severe facial pain. Definitive treatment consists of antiviral therapy and sometimes includes steroids.[8] The chances of recovery are better when definitive treatment is initiated within 3 days of the onset of the first symptoms.[9] In such instances, a 70% to 86% chance of full recovery can be expected, compared with an alarming 20% in untreated patients.[10] Prompt recognition of RHS is key to its successful management. Although facial swelling as one of the major presenting complaints is rare, it is reported here to help the dentist distinguish bacterial odontogenic infections from RHS and institute prompt antiviral therapy.

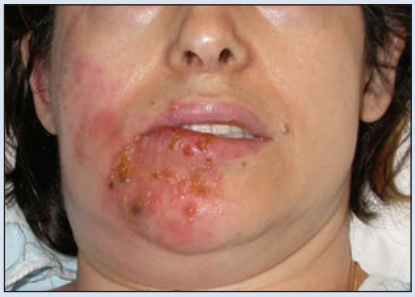

Figure 1a: Facial view showing unilateral vesicles, cellulitis, submandibular swelling and facial nerve paresis in the distribution of the buccal and marginal mandibular branches. Note the loss of definition of the ipsilateral nasolabial fold.

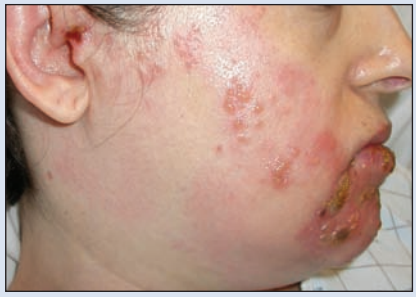

Figure 1b: Lateral view showing the initial extent and presentation of vesicles.

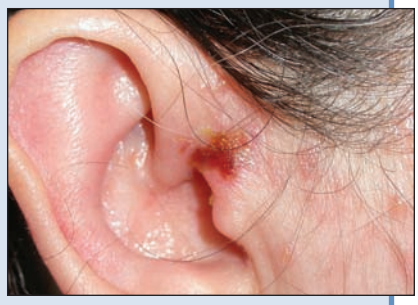

Figure 2: Initial crusted vesicles on the ipsilateral tragal region of the ear.

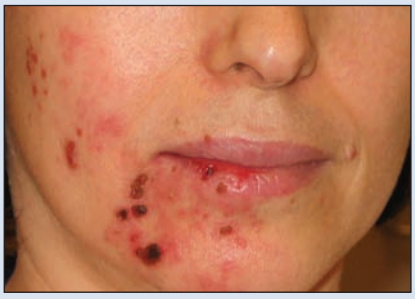

Figure 3: Black crusted and dried vesicles showing initial signs of resolution of the viral infection at 1-week follow-up.

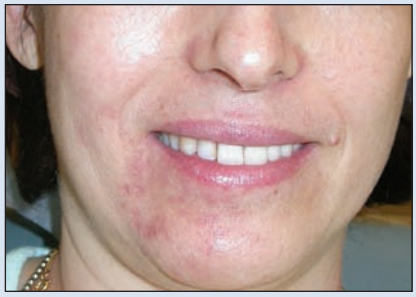

Figure 4: Resolution of marginal mandibular paresis is shown with return of the nasolabial fold on the right side. Note the healing perioral wounds at 1-month follow-up.

Case Report

A 43-year-old woman complaining of malaise, fever, facial pain and swelling was referred by an emergency physician for dental consultation regarding a suspected odontogenic infection. A 3-day history of fever had been followed by progressively worsening swelling, redness and generalized pain on the right side of the patient's face. An initial emergency consultation the day before at another hospital resulted in referral to a local dentist for treatment of an odontogenic infection. The patient's past medical history was significant only for a benign ovarian tumour, which had been surgically removed 6 months earlier.

Clinical examination revealed mild tachycardia, an elevated temperature of 39°C and lethargy. The facial swelling was centred in the right submandibular area, but extended to the cheek and parotid regions as well. These areas exhibited multiple erythematous, crusted vesicular eruptions, extending to the ipsilateral tragus, helix and pinna of the ear (**Figs. 1** and **2**). The patient stated that these vesicles were not present at her initial emergency room visit. A neurologic examination revealed mild weakness in the marginal mandibular branch of the ipsilateral facial nerve, which was best seen with a full smile (**Fig. 1a**). Loss of definition in the ipsilateral nasolabial fold also represented weakness in the buccal branch of the facial nerve. No other signs of neurologic impairment were noted. A comprehensive intraoral examination and dental radiographic examination resulted in the exclusion of an odontogenic source for the facial swelling and pain. The clinical signs and symptoms were diagnostic of RHS.

Intravenous fluid and analgesic therapy were provided while definitive antiviral treatment was instituted using acyclovir (400 mg orally, 4 times a day for 14 days). Additional antimicrobial therapy, consisting of cephalexin (500 mg orally, 4 times a day for 7 days), was also started as a prophylactic measure to prevent secondary bacterial infection of the facial skin lesions. No steroids were prescribed. After 1 week, most vesicles were crusted over and the facial paresis had resolved (**Fig. 3**). After 2 weeks, the patient was found to have a new dysesthetic and tingly sensation within the right tongue and lingual gingiva. However, all symptoms had resolved by 1 month (**Fig. 4**).

Discussion

VZV is a member of the Herpesviridae family. It has a lipid envelop surrounding the nucleocaspid and possesses a double strand of DNA with a total length of 125,000 base pairs.[7] This virus lies latent in the geniculate and other ganglia of the head and neck. RHS occurs in individuals who have previously had chickenpox and is caused by reactivation of VZV. It is characterized clinically by facial palsy and ipsilateral vesicular eruptions on the face and ears.[11,12] Other underlying systemic illnesses, particularly hematologic malignancies such as leukemia and lymphoma, may be associated with VZV reactivation.[13–16]

The facial paralysis seen in RHS is due to underlying neural inflammation, pressure and possible destruction of the facial nerve that takes place when reactivated VZV replicates within and travels along the axons of this nerve.[17] The facial nerve proper is principally a motor nerve. However, some sensory, parasympathetic and special sensory fibres run along its more proximal routes within the craniofacial skeleton. Specifically, nerves responsible for general sensation in parts of the ear, parasympathetic innervation of the pterygopalatine ganglion and special sensory taste in the tongue — which are all derived from the trigeminal, superior salivatory and solitary tract brainstem nuclei — join together in the brainstem to form the "nervus intermedius." The nervus intermedius in turn fuses with the facial nerve proper to enter the facial canal within the petrous temporal bone. The cell bodies of sensory components of the facial nerve are located within the geniculate ganglion. During the primary chickenpox infection, VZV travels along the nerve axons and eventually resides within this ganglion. During reactivation, inflammation and edema of the closely adjacent motor fibres result in paresis.[18] In this case report, the paresis was limited to those axons belonging to the marginal mandibular and buccal branches of the facial nerve. However, a significant facial swelling was also seen in the ipsilateral submandibular region. This may have been a result of edema due to concomitant viral reactivation within axons supplying the facial dermatome of the third division of the trigeminal nerve.

Acyclovir is an effective antimicrobial agent against actively replicating viruses. Acyclovir itself is not active. It must first be phosphorylated by viral thymidine kinase to form a triphosphate. Acyclovir triphosphate inhibits viral DNA polymerase and, thus, DNA replication.[19] Kinishi and others[8] showed that acyclovir therapy results in resolution of facial paresis and improved nerve function as measured by nerve excitability testing.[8] Although it is most effective, increasing viral resistance to acyclovir therapy has been reported[19,20] and newer drugs, such as valacyclovir, famciclovir, penciclovir and brivudine, are being used more commonly.[21–23] Adjunctive steroid therapy can be helpful in the management of the facial paralysis of RHS.[24] However, many authors caution against implementing steroid therapy, especially with periocular lesions, as they fear dissemination of the VZV infection.[24–26]

RHS may present with a spectrum of clinical variations, including facial swellings that appear to be of odontogenic origin. As a result, dentists may be challenged to make the correct diagnosis of RHS versus an odontogenic infection in a timely manner. Appropriate supportive and prompt antiviral therapy combined with close follow-up is associated with significantly better functional recovery and outcomes.[8]

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