CLINICAL VIGNETTE

Ramsay Hunt Syndrome (Type 2): An Atypical Presentation of a Very Common Disorder

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Case Report

The patient is a 62-year-old male with a history of GERD, hypertension, allergic rhinitis, and hypovitaminosis D who noted the acute onset of a burning pain in the left ear. Over the next two days, the symptoms spread into the left cheek and became much more intense. Three days later, he noted left-sided facial weakness and was seen by his ENT who noted an erythematous, vesicular rash in the left outer auricle and auditory canal. Cultures were taken and he was subsequently started on prednisoline and subsequently valaciclovir when the cultures grew herpes zoster. He was also given hydrocodone for pain control. MRI was ordered which showed mild generalized atrophy and scattered small subcortical white matter foci consistent with age. The left-sided facial weakness progressed over the next few days resulting in near-complete left facial paralysis and difficulty speaking. Audiology testing documented bilateral high-frequency sensorineural hearing loss greater on the left side in conjunction with the patient feeling a sensation of fullness and pain in the left ear without vertigo. Within the next week, the pain had improved but the left-sided facial paralysis was without change other than some slight improvement in his ability to close the left eye.

The patient was an alert, pleasant man, able to give a clear history. Vital signs included blood pressure 122/82, pulse 60 and regular, temperature 98.3 degrees Fahrenheit. A screening mental status evaluation was normal. He was oriented x3, and language was fluent. Overall cognitive function was intact. Recent and remote memory was normal. His gait was normal. He could stand in the Romberg position with eyes open and with eyes closed without difficulty. He could take 8 steps in tandem without a sidestep. Head and neck examination showed normal external auditory canals (the rash had resolved) and tympanic membranes bilaterally. Thyroid was normal to exam. On cranial nerve examination, extraocular movements were full in all directions without spontaneous or gaze-evoked nystagmus. Smooth pursuit and saccadic eye movements were normal. Head thrust testing showed normal symmetrical horizontal vestibular ocular reflex responses. Pupillary responses were symmetrical bilaterally. There was a near complete paralysis of the left side of the face. He could slightly move his upper lid downward and partially wrinkle his left forehead. There was no movement of the lower face on the left side. Lower cranial nerve function was normal. Motor examination showed normal muscle tone, strength, and bulk throughout. The deep tendon reflexes were normal bilaterally. Coordination examination showed normal rapid and rapid alternating movements without dysmetria or intention tremor. The screening sensory examination was normal.

Laboratory testing including comprehensive metablolic panel, complete blood count and ESR were within the normal ranges.

General Discussion and Historical Context

Ramsay Hunt Syndrome is a term used to describe three separate and distinct disorders all of which were described in 1907 by American neurologist James Ramsay Hunt (1872–1937). Dr. Hunt was born in Philadelphia in 1872 and graduated in 1893 from the University of Pennsylvania Medical School. He practiced neurology for many years in Manhattan with his research interests focusing on the anatomy and disorders of the corpus striatum and the extra-pyramidal system. He died in 1937[1].

Ramsay Hunt Syndrome Type 1 (Ramsay Hunt Cerebellar Syndrome) is very rare and causes degeneration of the cerebellum leading to ataxia, action myoclonus, tremor, and seizures. Ramsay Hunt Syndrome Type 3 (Hunt's Disease or Artisan's Palsy) is a neuropathy of the ulnar nerve. Ramsay Hunt Syndrome Type 2 (the most common type of Ramsay Hunt Syndrome and the topic of this paper) is a subtype of shingles involving reactivation of herpes zoster in the geniculate ganglion leading to

signs and symptoms that include facial nerve dysfunction, hearing loss, vertigo, tinnitus, nystagmus, and pain[1]. There is an acute peripheral facial neuropathy sometimes associated with the erythematous vesicular rash of the skin of the ear canal and auricle (also termed herpes zoster oticus), and/or mucous membrane of the oropharynx. Viral reactivation can lead to one or more of the following: herpes auricularis, herpes facialis, and herpes occipito-collairs. The neurons in the geniculate ganglion are responsible for the movements of facial muscles, sensation of the ear and ear canal, the taste of the frontal two-thirds of the tongue, and the moisturization of the eyes and the mouth. While most cases of shingles involve primarily sensory symptoms, Ramsay Hunt typically affects both motor and sensory function[2]. When it occurs in the absence of a skin rash, it is known as zoster sine herpete[3].

Epidemiology

Ramsay Hunt Syndrome has historically been considered a relatively uncommon form of shingles seen almost exclusively in the elderly population. However, it is now know that about 18% of all facial palsies are related to Ramsay Hunt[4]. Moreover, recent reports suggest that it may be the cause of about 20% of cause of Bell's palsy[5]. Interestingly, varicella virus has been detected by polymerase chain reaction (PCR) in the tear fluid of some patients diagnosed with Bell's palsy[4]. As in all types of zoster reactivation, the incidence and severity of Ramsay Hunt increases with age although rare cases have been reported in children[6]. The increase in incidence and severity with age is believed to relate to changes in cellular rather than humoral immunity[7]. Immunocompromised patients are more likely to develop Ramsay Hunt and are more likely to have varicella reactivation in multiple dermatomes[8].

Etiology

Ramsay Hunt Syndrome is caused by reactivation of the varicella zoster virus in the geniculate ganglion of the brain.

Clinical Features

The most common presenting symptom is pain deep within the ear. This pain often fluctuates for a day or two, then becoming more constant and severe. The classic shingles skin findings (an erythematous vesicular rash) often appears in the ear canal and auricle hours to days later sometimes resulting in secondary infections. During the first week, other symptoms such as vertigo, hearing loss, tinnitus, and a facial droop may become apparent[9]. Affected areas tend to include the anterior two thirds of the tongue and the soft palate[5]. The unilateral facial nerve dysfunction may be clearly seen on examination or may be elicited on testing. Hyperacusis is sometimes seen due to reduced nerve function to the stapedius and tensor tympani[1]. Rather severe ataxia may also be a prominent finding. The muscles of the forehead are usually spared as innervation of the forehead is bilateral. Rarely will other cranial nerves (5, 6, 8, 9, and 10) be involved[1]. Facial nerve dysfunction usually is most severe by day seven with ipsalateral hearing loss reported in about 50% of cases[9].

Diagnosis and Testing

Ramsay Hunt is very similar clinically to Bell's palsy except that the characteristic varicella rash is not seen in a pure Bell's palsy. Making the diagnosis more difficult is the fact that some patients (especially younger patients) may only have ear pain without a noticeable rash. Differential diagnosis includes CVA, viral labyrinthitis, trigeminal neuralgia, temporomandibular disorders, dental abscesses, otitis media, and otitis externa[10].

In terms of testing, virological studies are available (typically by PCR of vesicle fluid) but usually the diagnosis is made on clinical grounds[11]. Varicella culture isolation is often time consuming and has a low sensitivity while PCR, antigen detection by DFA, and Tzanck staining are generally more rapid and sensitive[12]. Audiometry may be performed to document the extent of associated hearing loss. Occasionally, nerve conduction studies may be done to determine the damage to the facial nerve and potential for recovery. WBC count, erythrocyte sedimentation rate (ESR), and serum electrolytes are sometimes helpful in assessing the infectious and inflammatory nature of this syndrome. Spinal fluid analysis and CNS imaging studies are occasionally recommended when CNS complications (such as meningitis, encephalitis, and vasculitis) are suspected[13]. Imaging studies such as MRI, CT scan, and MRA can be useful to rule out structural lesions[14].

Several grading scales have been developed to quantify the severity of Ramney Hunt Syndrome with the House-Brackmann facial neuropathy scale being

commonly used[5]. The House-Brackmann facial neuropathy scale includes[5]:

- 1 - Normal
- 2 - Mild dysfunction (slight weakness only noticeable on close inspection)
- 3 - Moderate dysfunction (obvious weakness, but not disfiguring differences between both sides)
- 4 - Moderately severe dysfunction (obvious weakness and disfigurement)
- 5 - Only barely perceptive motor function
- 6 - Complete paralysis

Treatment

The most commonly used pharmacologic treatments are antiviral agents and corticosteroids. A recent study, however, showed that adding intravenous acyclovir to corticosteroids did not improve outcomes after six months[15]. Another recent study concluded that controlled-release oxycodone was generally effective in patients experiencing acute pain due to herpes zoster[16]. Carbamazepine and gabapentin may also be helpful in some cases[16]. As with Bell palsy, care must be taken to prevent corneal irritation and injury. Surgical options do not have a role in the management of this disorder unless a structural lesion is found on imaging studies.

Prognosis

Poor prognostic factors tend to include an age older than 50 years, diabetes, hypertension, complete facial paralysis, and lack of 7th cranial nerve excitability on examination[2]. Early diagnosis and initiation of treatment (within 72 hours of the onset of symptoms) is associated with improved outcomes[1]. 75% of patients had a complete recovery when therapy was initiated within 72 hours of onset of symptoms while treatment is still beneficial for up to one week after the appearance of symptoms[17]. Partly because some patients do not receive medical therapy within the first week, fewer than 50% of patients have a full recovery of nerve function[2]. Medications don't appear to have much of a role in the recovery of any hearing deficits[17]. Prevention of Ramsay Hunt Syndrome is based on appropriate vaccination with the zoster vaccine. Ramsay Hunt Syndrome is typically a self-limited condition with a fairly good prognosis. Morbidity tends to be related to hearing loss, tinnitus, vertigo, and facial nerve palsy. Ramsay Hunt patients are less likely to fully recover than patients with Bell's palsy[18].

Clinical Course and Follow-Up

The patient was immediately started on anti-viral and corticosteroid medications. MRI imaging of the brain was negative as was his bloodwork. He was seen by his neurologist at 2 week, 6 week, and 3 months intervals and was noted to have steady improvement. By 6 months, he was about 95% back to his baseline state with just a small residual facial nerve dysfunction which was barely noticeable.

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Submitted on February 21, 2013