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Case report

Continuous hemicrania as the initial manifestation of Ramsay Hunt syndrome: a case report

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Abstract

Introduction
Ramsay-Hunt syndrome, also called otic zoster, is a rare complication of herpes zoster. The syndrome is characterized by peripheral facial nerve palsy and an erythematous vesicular eruption in the ear or mouth. Preceding the appearance of the vesicles, unilateral otalgia or neck pain may occur more commonly. However, persistent hemicrania is infrequent in the preeruptive phase.

Objective
To present an atypical case of Ramsay Hunt syndrome with continuous unilateral headache preceding the onset of other symptoms and signs of the syndrome.

Case report
Report of a 69-year-old woman who presented subacute onset of moderate to severe left hemicrania with no autonomic signs. Eight days after the start and continuous headache maintenance, she presented with peripheral facial paralysis. After four days, she noticed the presence of vesicles in the left ear and odynophagia. She developed nausea with several episodes of vomiting and severe imbalance that made it impossible for her to walk unassisted. On physical examination, she presented vesicles in the left ear and oropharynx, left peripheral facial palsy (House Brackmann grade IV), left hypoacusis, nystagmus, and vestibular gait. Diagnostic tests for screening several metabolic diseases and diagnosis of infection (including HIV) were unremarkable. Brain computed tomography and cerebrospinal fluid analysis showed no abnormalities.

Conclusion
Ramsay-Hunt syndrome mainly involves the facial and vestibulocochlear nerves, causing peripheral facial palsy, otalgia, hypoacusis, and, less frequently, imbalance. Although pain is a frequent manifestation of the preeruptive phase of RHS, unilateral headache is not common in this scenario. On the other hand, it is a prevalent complaint in the emergency department and has several different etiologies. Hence, diagnosing RHS when patients present exclusively unilateral headaches is challenging for clinicians. Physicians must consider RHS a vital differential diagnosis of sided-locked headaches, avoiding diagnostic errors and treatment delays.

Keywords:
Herpes zoster oticus
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Introduction

Ramsay Hunt syndrome (RHS) is a rare condition characterized by the reactivation of the Varicella Zoster virus in the geniculate ganglion of the seventh cranial nerve.[1] The first contact with the virus results in fever and a diffuse vesicular eruption known as chickenpox. After the initial infection, the virus remains latent within various ganglia and can be reactivated at any point. This reactivation is called herpes zoster (HZ) or geniculate herpes zoster (or herpes zoster oticus) when it occurs in the facial nerve.

The clinical diagnosis encompasses the triad of ipsilateral facial palsy, otalgia, and vesicles near the ear and auditory canal. Other manifestations include a change in taste, dry eye, hyperacusis, nasal obstruction, hoarseness, aspiration, hearing loss, tinnitus, nystagmus, vertigo, nausea, and vomiting, depending on the involvement of other nerves.[2] The most affected cranial nerves besides VII are VIII and X, and the involvement of IX and XII is less reported.[3]

Neurological symptoms usually appear before cutaneous manifestations, and initially, the syndrome can be confused with other neurological conditions that cause facial paralysis. Severe ear pain is the most common initial symptom, followed by paralysis.[4] Epidemiological data indicate that the syndrome is the cause of 7% of facial paralysis.[5] Furthermore, it can occur at any age, but the elderly are more susceptible to developing HZ.[6]

Early diagnosis is crucial, as is the initiation of treatment aimed at reducing the duration of the disease, providing analgesia, and preventing complications. A delay in diagnosis can result in increased long-term complications. Complications include synkinesia, permanent facial paralysis, corneal abrasions and ulcers, postherpetic neuralgia, hearing loss, and tinnitus.[7] Additionally, late complications of RHS, such as cerebral venous thrombosis, have recently been described.[8]

Mainly, it is highly challenging for clinicians to diagnose RHS when patients complain only of headaches, without facial weakness, dizziness, or typical vesicles in the ear.

Case report

A 69-year-old woman complained of subacute onset of a daily stabbing left-sided headache of moderate to severe intensity. Her headache was always of left frontotemporal distribution, and it would never move to the right side. She experienced periods of pain exacerbation, lasting some hours when she sought the emergency room. During these periods, she would develop dizziness, nausea, and vomiting; she could not function at her job and had trouble completing any of her activities of daily living. She did not complain of nasal congestion or rhinorrhea and denied eyelid droop, lacrimation, or eye redness. She sought several medical services and was prescribed several analgesics without improving her symptoms. She had no significant headache history until 12 days prior when her headache started as daily from the onset. Eight days after the beginning of the headache, she developed peripheral facial paralysis, and after four days, she noted the appearance of vesicles on the left ear and odynophagia. She developed nausea with several episodes of vomiting, and severe imbalance made her unable to walk straight. Shortly after, the patient was admitted to our institution

On physical examination, she was conscious, oriented, and afebrile. She presented with vesicles in the oropharynx and left ear (Figure 1), left peripheral facial paralysis House Brackmann grade IV (Figure 2), left hypoacusis, nystagmus, and vestibular gait. The results of routine hematologic and blood-chemistry studies were normal, as were the values for vitamin B12, folic acid, and thyroid-stimulating hormone. Serology for B and C hepatitis, and HIV were negative. Brain computed tomography without and with contrast and analysis of the cerebrospinal fluid showed no abnormalities. Intravenous acyclovir and oral prednisone were prescribed, with good clinical evolution. The patient signed informed consent and authorized the publication of images, including her face and ear.

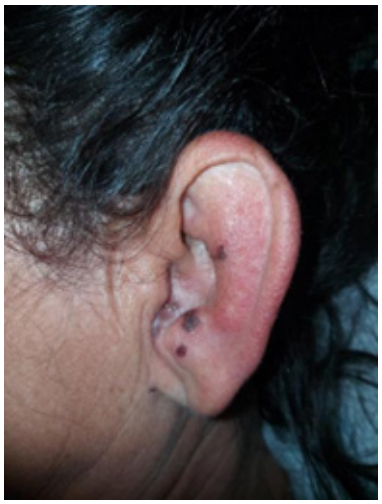

Figure 1. Vesicles in the left ear.

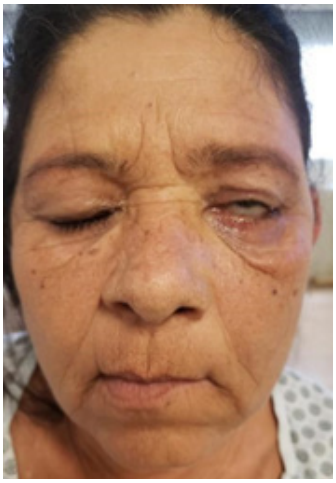

Figure 2. Left peripheral facial paralysis.

Discussion

The most challenging point of this case is the initial manifestation of RHS as a continuous unilateral headache without autonomic signals. Side-locked primary headaches are commonly associated with a short duration of attack (less than 4 hours). This group includes trigeminal autonomic cephalalgias (TAC).[9]

Still, less habitually, long-lasting headaches may likewise present with the pain always on the same side, including migraine, hemicrania continua, and cervicogenic headache.[10, 11]

Besides side-locked unilaterality, the acute onset and the continuous headache pattern reported here deserve more consideration.

Epidemiology of sided locked headache with the acute or sub-acute onset

Prakash et al.[10] prospectively evaluated 307 consecutive adult patients presenting to a neurology outpatient clinic in India with side-locked unilateral headaches and facial pain. They found that only 8% of their patient series had a headache history of fewer than three months. The authors highlighted some very interesting points: primary headaches account for only 19% of patients in this group; a daily or continuous headache pattern was clearly associated with either secondary headaches or headaches included in group 13 of ICHD-39 a significant number of patients (19%) could not be classified in the acute stage. All patients of this subgroup complained of daily headaches since the start. They pointed out that some patients may have secondary headaches in a nascent stage, where other clinical manifestations or abnormalities would emerge later, similar to our patient. In another specialized clinic-based study, Ramón et al.[11] identified 13 secondary headaches among 100 strictly unilateral headache consecutive patients; this represents the second most prevalent headache diagnosis of their series. They reported the following causes: two cases of herpetic infection of the occipital nerve, dissecting aneurysms of the vertebral artery and temporal arteritis, and one case secondary to parietal meningioma, arachnoid cyst, airplane travel, orbital myositis, Tolosa-Hunt syndrome, Chiari type I malformation and skull injury.

Epidemiology of sided locked headache with a continuous pattern

When a physician deals with a patient suffering from a unilateral continuous headache, an initial diagnostic hypothesis could be hemicrania continua (HC). HC is a one-sided headache that is always present and is usually of mild to moderate intensity, with pain exacerbations periods of severe headache with accompanying autonomic and/or migrainous features.[9,12] Nevertheless, the ICHD-3[9] criteria require a headache presence for >3 months and an absolute response to therapeutic doses of indomethacin.[9,13] Some disorders may mimic HC, such as intracranial space-occupying lesions, posttraumatic headache, post-craniotomy headache, post-stroke headache, internal carotid artery (ICA) dissection, or aneurysm, analgesic rebound headache, idiopathic intracranial hypertension, venous malformation, cerebral venous sinus thrombosis, paraneoplastic syndrome, sinus pathologies, dental lesions, and temporomandibular joint diseases.[14]

Two other differential diagnoses one can speculate in this clinical scenario are the "New Daily Persistent Headache" (NDPH)[15] and Cervicogenic Headache (CGH).9 NDPH characteristically presents with a sudden onset headache, which starts and remains without remission, but a bilateral headache typifies this condition. Accordingly, the diagnosis of NDPH was made in none of the patients evaluated by Ramón et al.[11] and in only 3% by Prakash et al.[10] CGH presents as unilateral pain that starts in the neck, and its prevalence was 8.1% among the side-locked headaches.[10] CGH, in clinical practice, is a unilateral (side-locked) non-throbbing headache that starts in the posterior part of the head and neck and extends to the head's ipsilateral side, occasionally reaching the ocular region. Sometimes the frequency could upturn, and a daily or near-daily headache develops.[16] Additionally, the demonstration of a cervical cause is at the core of the diagnosis.[9]

Association of acute herpes zoster of the head and headache or facial pain

A frequent complaint of patients with Bell's palsy is ipsilateral periauricular pain that starts days before the onset of the paralysis.[17] Bell palsy occurs most often in pregnant women and patients with diabetes mellitus, hypertension, and infection by some types of virus, such as influenza, herpes simplex, and herpes zoster. Herpes zoster may involve the trigeminal ganglion in about 10% of cases, and the first division of the trigeminal nerve is singled out in some 80% of patients.[9] Zoster sine herpete (ZSH) is an unusual manifestation of VZV infection and presents with similar symptoms but without a vesicular rash. Polymerase chain reaction detection of varicella-zoster virus DNA in the cerebrospinal fluid may diagnose such cases.[18] HZ, or shingles, is the eruption of a rash, typically unilateral and painful, caused by the VZV. During this preeruptive phase, about 70–80% of patients with HZ experienced dermatomal pain and healing of the skin, with the resolution of pain occurring within three weeks of rash onset.[19] Lee et al.[20] retrospectively studied the clinical features of 152 patients with acute HZ infection restricted to the head who presented within ten days of rash onset at a tertiary hospital in South Korea between January 2011 and December 2015. They found that preeruptive pain was reported by 112 of the 152 patients, with a mean duration of 3.0±1.3 days (range, 1–6 days). The pain was characteristically felt in the nerve's territory, and only 27.6% of the cases present an ipsilateral spread beyond the nerve's territory. These attacks are moderate to severe in intensity and throbbing in character. The authors advocate that a headache that is stabbing in quality, primary or secondary, is a crucial differential diagnosis of the preeruptive pain of acute HZ of the head.[20] Interestingly, some cases of short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) related to the infection of VHZ have been reported.[21-23] A thorough overview of clinical aspects of painful trigeminal neuropathy attributed to HZ[9] is beyond the scope of this article and has been reviewed in detail elsewhere.[24]

Ramsay Hunt Syndrome

Ramsay Hunt syndrome was foremost described in 1907 by James Ramsay Hunt, representing an essential differential diagnosis of Bell's palsy.[1] Other conditions reported as differential diagnoses include primary chickenpox, stroke, poison ivy, cellulitis, herpes simplex virus, and trigeminal neuralgia.[7] The clinical manifestations of HZ will depend on the nerves involved, and less than 1% involve the facial nerve.1-3 The clinical diagnosis of SRH is straightforward and based on anamnesis and neurological examination. Preceding the appearance of the vesicles in RHS, unilateral otalgia or neck pain may occur more frequently.[1,7] Unilateral headache is not a frequent manifestation of the preeruptive phase of VZV in RHS, as presented by our patient.

Compared with Bell's palsy, patients with Ramsay-Hunt syndrome have a worse long-term prognosis and are less likely to recover fully.[4,5] The most important prognostic indicator in Ramsay Hunt syndrome is the severity of the facial palsy, although age above 50 is also associated with worse outcomes.[3] The most effective treatment for long-term complications is antivirals associated with corticosteroids if administered early.[1,7]

Conclusion

When facing a patient with one-sided headaches, particularly always present, the physician must think about a secondary cause of the head pain. In this scenario, RHS is an essential diagnosis to consider, even when the typical clinical picture is absent. Hence, suspicion, early diagnosis, and correct management are crucial to provide the patient better prognosis and to keep away debilitating complications as much as possible.

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