Case Report

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Ramsay Hunt syndrome affecting the vagus nerve with epiglottic ulcers as the first manifestation: a case report

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Abstract

Ramsay Hunt syndrome involving the vagus nerve is very rare. We herein describe a 53-year-old man who developed severe pharyngeal pain after alcohol intoxication. Antibiotic treatment was ineffective. Laryngoscopy showed an ulcer on the right side of the epiglottis. As the condition progressed, the patient developed hoarseness. He then gradually developed multiple herpes lesions on the right side of the head, face, and neck along with right peripheral facial paralysis. Corticosteroid, analgesic, antiviral, and nutritional nerve therapy resulted in disappearance of the herpes lesions, epiglottis ulcer, pharyngeal pain, and right head and face pain. The facial paralysis slightly improved, but the hoarseness did not improve. The patient was discharged with an unsatisfactory outcome, and he attempted treatment with acupuncture. After 6 months, the right facial paralysis and hoarseness disappeared. Our case emphasizes the importance of early diagnosis and treatment of atypical Ramsay Hunt syndrome as well as timely communication, enhancement of trust, and reduction of disputes between doctors and patients.

Keywords

Vagus nerve, epiglottis, ulcer, herpes zoster, varicella-zoster virus, Ramsay Hunt syndrome, facial paralysis

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Introduction

Ramsay Hunt syndrome is caused by varicella-zoster virus (VZV), the causative virus of herpes zoster. Ramsay Hunt syndrome is a complication of herpes zoster in which VZV invades the facial nerve in the head and neck, causing facial paralysis.

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However, invasion of the vagus nerve by VZV is very rare. Cao et al.[1] reported a case of VZV encephalitis with glossopharyngeal and vagus nerve injury as the primary manifestation combined with a medulla lesion. In that case, the patient exhibited dysphagia and a hoarse voice and was misdiagnosed with lateral medullary syndrome. Gunbey et al.[2] described a patient who was infected by VZV with cranial nerve VII, IX, and X involvement as shown by magnetic resonance imaging. Unlike our patient, the patient described by Gunbey et al.[2] initially presented with Ramsay Hunt syndrome. We herein report a case of Ramsay Hunt syndrome with epiglottic ulcers as the first manifestation to help clinicians better understand this atypical presentation of Ramsay Hunt syndrome.

Case report

A 53-year-old man with a 2-day history of odynophagia and dysphagia was admitted to our hospital. He had a cough but no fever, dyspnea, or hoarseness. After 2 days of oral antibiotic treatment outside the hospital, his symptoms did not improve. The patient had become heavily intoxicated by alcohol 2 days before presentation. On his first day as an inpatient, laryngoscopy revealed an ulcer in the right side of the epiglottis, but his bilateral vocal cords moved well without swelling. After admission, he received antibiotic therapy (amoxicillin/potassium clavulanate at 1.2 g intravenously three times a day) to prevent infection. He was hospitalized on the second day after admission. Routine blood parameters, urinalysis, infection markers, tumor markers, a fungal D-glucose test, and procalcitonin level were normal. On the fourth day after admission, the patient's pharyngalgia became aggravated and hoarseness appeared. Physical examination showed that the right vocal cord was fixed, and ulcers were found on the right side of the epiglottis, aryepiglottic fold, and soft palate (Figure 1(a) and (b)). No space-occupying lesion or bone destruction was found on laryngeal computed tomography. No compression of the recurrent laryngeal nerve was found on chest computed tomography. The patient was treated with acyclovir (0.25 g intravenously three times a day), a corticosteroid (methylprednisolone at 80 mg intravenously once a day), and nutritional neurotherapy. On the seventh day, the patient's pharyngalgia had improved, but his hoarseness remained unchanged. Herpes lesions were found on the right neck, right auricle, and temporal region with marked pain. On the eighth day, the patient's pharyngalgia continued to improve, his swallowing function improved,

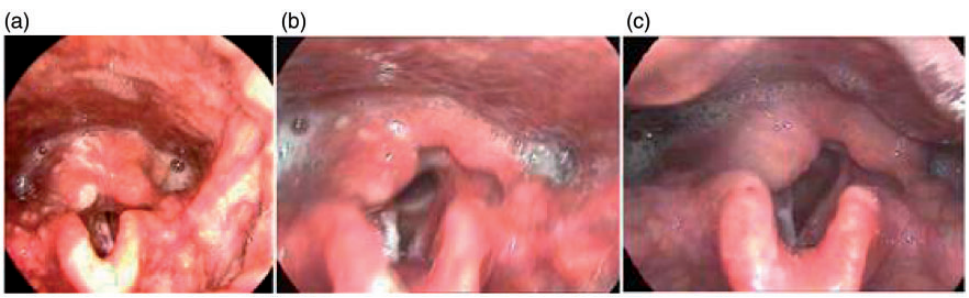

Figure 1. Physical examination findings. (a, b) The right vocal cord was fixed, and ulcers were found on the right side of the epiglottis and aryepiglottic fold. (c) The ulcers disappeared, but effusion in the right pyriform sinus remained and the right vocal cord was still fixed.

and his throat ulcers decreased in severity. On the 14th day after admission, the patient's pharyngalgia disappeared and his swallowing function improved, but his voice was still hoarse. Physical examination showed that the ulcers had disappeared but that right pyriform sinus effusion remained and the right vocal cord was still fixed (Figure 1(c)). The herpes lesions on the right neck, auricle, and temporal region disappeared. However, the right frontal striae became shallow without movement, the right eye could only be completely closed with strong force, the right nasolabial groove became shallow, and the corner of the mouth slanted to the left when speaking. No skew at the mouth corner was present in the static state. According to the House–Brackmann facial nerve grading system,[3] the patient had grade IV facial paralysis. Because of the poor therapeutic effect and positivity of VZV antibody, we added interferon for antiviral treatment (interferon at 3 million units intramuscularly once every other day). On the 20th day, the symptoms of right facial paralysis improved, but the hoarseness remained unchanged. Obvious effusion was present in the right pyriform fossa. The right frontal striae had become deeper and could move, and the right eye could be closed completely with slight force. The patient's facial paralysis had improved to grade II.

The patient was not satisfied with the therapeutic effect and was discharged without medical advice. He chose acupuncture as a rehabilitation treatment for 1 month. After 6 months, the patient's facial paralysis and hoarseness gradually disappeared. Laryngoscopy showed that the right vocal cord had resumed normal movement.

Discussion

Ramsay Hunt syndrome is caused by VZV in the geniculate ganglia of the facial nerve. It affects multiple cranial nerves but mainly involves the facial nerve and auditory nerve. James Ramsay Hunt first reported this condition in 1907.[4] VZV, which widely exists in China, is common in adults, the elderly, and patients with immunodeficiency. Through an epidemiological survey of people aged >50 years in Guangdong Province, Zhu et al.[5] found that the lifetime incidence rate of herpes zoster is 3.46%. To date, no epidemiological investigation of VZV has been conducted in China. Herpes zoster is caused by reactivation of the pre-existing virus. The main manifestations are abnormal skin sensation, pruritus, pain, and finally herpes lesions. The lesions are connected in series into a band, mostly seen on the trunk and forehead, and distributed on one side. Throat discomfort caused by VZV infection of the glossopharyngeal nerve and vagus nerve is very rare and is easily missed and misdiagnosed. The patient in our case had been severely intoxicated by alcohol 2 days before symptom onset.

Herpes zoster should be considered when a patient shows multiple unilateral ulcers in the throat and antibiotic treatment is ineffective. The disease often progresses rapidly during the treatment of such patients. Early and accurate judgment of the disease is helpful for communication between doctors and patients and gains the trust of patients. This is particularly important in today's tense relationship between doctors and patients in China.[6] When a classified diagnosis and treatment are lacking, most patients go directly to a tertiary hospital in the beginning of their clinical course. If the patients do not attain a good curative effect, the doctor can easily lose the patient's trust. Our patient had been hospitalized for 20 days. We had informed the patient in advance about the possible symptoms and signs and had thus gained his trust. Combined treatment with acyclovir and a corticosteroid can reportedly hasten

the recovery of facial palsy.[7] Although we administered acyclovir and corticosteroid combination therapy for a long period of time, the outcome was still unsatisfactory. The patient was dissatisfied with the effect and finally discharged without medical advice.

The diagnosis of Ramsay Hunt syndrome is based on auricular pain, herpes in the ear, and peripheral facial paralysis of different degrees on the same side. According to the patient's history, presence of ear herpes, and other factors, the clinical diagnosis is usually not difficult. In our patient, however, the early throat ulcers (or herpes mucosa) were not easy to identify. Ramsay Hunt syndrome is mainly differentiated from Bell's facial paralysis. Herpes zoster in the throat should be differentiated from tuberculosis, syphilis, and Fanshang's angina. Pharyngeal and laryngeal VZV infection manifests as scattered ulcers in the corresponding nerve distribution area. The ulcers are mostly unilateral and distributed along the nerve, and they generally do not cross the midline. The cause of vocal cord paralysis should be differentiated from brain nerve injury and space-occupying lesions. Herpes zoster paralysis has a worse prognosis than Bell's palsy with respect to recovery of facial function, even with a high dose of corticosteroids.[8]

Because of the presence of a possible viral etiology and poor facial function outcomes, some investigators have pointed out that the use of acyclovir provided better results in some preliminary reports.[9,10] Early and sufficient use of antiviral drugs and corticosteroids is more effective than use of only corticosteroids in terms of avoiding complications, such as permanent facial nerve dysfunction.[7,11,12] Although many studies have shown that interferon can inhibit VZV, the use of interferon in Ramsay Hunt syndrome is still controversial. Interferon has not been mentioned in the Chinese or European guidelines for the treatment of Ramsay Hunt syndrome.[13,14] The recovery of cranial nerve function often takes a long time. Even if the curative effect is not good, long-term rehabilitation therapy is also of great significance. Acupuncture in traditional Chinese medicine plays a beneficial role in this kind of rehabilitation treatment.[15]

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics

Because this was not a clinical trial, ethics approval was unnecessary. Informed consent was obtained from the patient.

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Supplemental Material

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