



Ramsay-Hunt Syndrome with Maxillary Dermatome Involvement: A Case Report

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Abstract

Herpes zoster (HZ) is caused by the reactivation of varicella zoster virus (VZV) that remains latent in the dorsal root ganglia after varicella infection. During the course of the disease, vesicular eruptions appear on skin of the innervated dermatome. Herpes zoster usually occurs in elderly patients. Thoracic dermatome is the most involved area in HZ. Trigeminal nerve is the most affected cranial nerve and ophthalmic branch of the trigeminal nerve involvement (ophthalmic zona) is twenty times more frequent in comparison with involvement of maxillary and mandibular branches. Ramsay-Hunt syndrome is a cranial polyneuropathy following VZV reactivation. The frequency of Ramsay-Hunt syndrome is defined as %1 in HZ infections. We present this interesting case because maxillary branch involvement with Ramsay-Hunt syndrome is a rare combination.

Key Words: Ramsay Hunt Syndrome; Herpes Zoster; Maxillary Zona Zoster.

Maksiller Dermatomu Tutan Bir Ramsay-Hunt Sendromu Olgusu

Özet

Herpes zoster (HZ) varisella enfeksiyonu sonrası arka kök gangliyonlarına yerleşen varisella zoster virüsünün (VZV) reaktivasyonu sonucu, latent kaldığı gangliyonun innerve ettiği dermatom bölgesinde veziküler lezyonlarla ortaya çıkar. Sıklıkla yaşamın ilerleyen dönemlerinde görülür. Zonada en sık tutulum torasik dermatomda olmaktadır. Kraniyal sinir tutulumu ise en sık olarak trigeminal sinirde olup, oftalmik dalın etkilenmesi (oftalmik zona), maksiller ve mandibuler dal tutulumuna göre yaklaşık yirmi kat daha siktir. Ramsay-Hunt sendromu, varisella zoster virüsünün reaktivasyonu ile ortaya çıkan bir kraniyal polinöropatidir. Herpes zoster enfeksiyonları içerisindeki sıklığı %1 olarak tanımlanmıştır. Maksiller dal tutulumunun nadir olması ve beraberinde Ramsay-Hunt sendromu bulunması yönüyle enteresan bulunan hastanın sunulması uygun görülmüştür.

Anahtar Kelime: Ramsay Hunt Sendromu; Herpes Zoster; Maxiller Zona Zoster.

INTRODUCTION

Herpes zoster often occurs in later stages of life due to the reactivation of the virus settled in the dorsal root ganglia after varicella and it comes out with vesicular lesions in the dermatome region innervated by the ganglion where it remains latent (1). It equally affects both sexes and all races (2). The typically affected zones in shingles are reported to be thoracic (53%), cervical (20%), ophthalmic (15%), and lumbosacral (11%) regions (3). Approximately 20 times more common than ophthalmic branch zoster (ophthalmic zona) and maxillary and mandibular branch involvement, cranial nerve involvement is most common in the trigeminal nerve (4).

Herpes zoster involves cutaneous (bacterial superinfection, scarring, zoster gangrenosum, cutaneous spread), visceral (pneumonia, hepatitis, esophagitis, gastritis, pericarditis, cystitis, arthritis), and neurological (postherpetic neuralgia, meningoencephalitis, transverse myelitis, peripheral nerve palsy, cranial nerve palsies, sensory loss, eye complications, deafness) complications (5). Ramsay-Hunt syndrome, on the other hand, is a

cranial polyneuropathy characterised by herpetic eruptions with facial nerve involvement and auricular, and sometimes oropharynx, involvement (6). Ramsay-Hunt syndrome has an incidence rate of 1% among other Herpes zoster infections (7).

CASE REPORT

A male patient around his seventies presented with rash, pain, and swelling in his face that had started a day before his admission to our clinic. Dermatological examination showed a viable erythema, distinctive oedema, and temperature rise in the left half of his face. We observed hemorrhagic scabs and millimetric vesicles here and there on the left cheek, on the front side of the ear, and around the auricle (Figure 1). On the left half of the tongue and the buccal mucosa, we observed millimetric ulcers and pustules (Figure 2). The systemic examination showed no pathologies. We applied Tzanck smear test, which is used to detect herpes virus infections as well as in differential diagnosis of vesiculobullous diseases. The test result was positive for our patient. With a diagnosis of herpes zoster, the patient was referred to the Dermatology service for his hospital stay. Due to poor oral intake, we started a

parenteral acyclovir treatment. Having severe ear pain on the second day of his hospitalisation, the patient was consulted to otorhinolaryngology clinic. Once at the otorhinolaryngology clinic, our colleagues detected moderate peripheral facial paralysis (Grade-4) and the patient was diagnosed with Ramsay-Hunt syndrome. Therefore we started an intravenous administration of prednisolone 60 mg/day. Responding to the treatment and his erythema decreasing, the patient was diagnosed with Ramsay-Hunt syndrome accompanied by maxillary involvement of shingles due to the anatomical location of the lesions. The patient was administered acyclovir for 7 days. In the end, his steroid dose was reduced gradually. The treatment brought about regression in the skin lesions and near complete recovery of facial functions.



Figure 1. Hemorrhagic scabs and millimetric vesicles on the left cheek, on the front side of the ear, and around the auricle.



Figure 2. Millimetric ulcers and pustules on the left half of the tongue and the buccal mucosa.

DISCUSSION

Herpes zoster is an acute and painful dermatitis due to the reactivation of varicella zoster virus in the dermatome. Following a VZV incidence, the primary cause of infection in varicella, the virus, which has latently remained in the dorsal root or trigeminal ganglia, is reactivated when the host resistance drops

below critical levels and eventually causes HZ lesions characterised by painful vesicles in the dermatome that is innervated by the ganglion (8).

Febrile illnesses, fatigue, intense stress, traumas, treatment with cytotoxic drugs, immunosuppression, exposure to radiation, malignancies, old age, alcoholism, and dental manipulations are among preparatory factors for HZ (9). The most important risk factor in the emergence of the lesion, however, is old age (10). In our patient, we could not find any underlying causes for his current state and so, we concluded that his condition was a result of his old age. Among the cranial nerves, it is the trigeminal nerve that gets involved in such conditions the most (4). In our patient, the involvement was limited to maxillary branches. In the maxillary and mandibular involvement of the trigeminal nerve, ulcers are seen on the tongue, palate, and floor of mouth (11). About 30 days after the zoster due to similar involvements, maxillary and mandibular alveolar bone necrosis may develop (5). In the case of our patient, we observed ulcers on the tongue and palate though there were not any skeletal complications. As one of the neurological complications of herpes zoster, Ramsay-Hunt syndrome is a cranial polyneuropathy following the reactivation of VZV (6). Although it basically affects VIth cranial nerve, it can also take place due to the involvement of cervical plexus, which is made of Vth, IXth, and Xth cranial nerves, which are connected to facial nerves, and IInd, IIIrd, and IVth cervical nerves (12).

Chang et al.'s study notes that 11.8% of their HZ patients had neurological complications. In their study, the most common complications were Ramsay-Hunt syndrome and segmental paresis of extremities (13). The incidence of Ramsay-Hunt syndrome among HZ in infections has been reported as 1% (7). In our patient, who had maxillary zoster, we observed the development of facial paralysis about 2 days after his hospitalisation, which helped us conclude that he had Ramsay-Hunt syndrome.

Starting the administration of antiviral agents such as acyclovir, valaciclovir, and famciclovir particularly within the first 72 hours is important in terms of the effectiveness of the treatment (14). Murakami et al. have reported to have used acyclovir together with prednisone in the treatment of Ramsay-Hunt-syndrome related facial paralysis and have stated that they had a remission rate of 75% in patients who started to take the medication within the first 3 days and a remission rate of 30% in patients who started to take the medication after the 7th day of the paralysis (15). We started the medication within the first 3 days and observed a near complete recovery of facial functions.

To sum up, due to the anatomical location of the lesions, we detected maxillary shingles followed by a diagnosis of Ramsay-Hunt syndrome in our patient's case. Because maxillary involvement along with the development of Ramsay-Hunt syndrome is a quite uncommon condition, we regarded our patient to be a noteworthy case to publish.

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