Ramsay Hunt Syndrome in Two Children: Case Report

İki Çocukta Ramsay Hunt Sendromu

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ABSTRACT Cranial facial nerve palsy is uncommon in childhood. Diagnostic studies in children and adults have provided convincing evidence that reactivation of varicella-zoster virus (VZV) may lead to cranial nerve VII palsy, which is the most common cause of facial palsy in childhood. Careful physical examination especially detailed ear examination is very important when VZV reactivation is suspected. Early treatment with acylovir therapy may improve the recovery rate of facial nerve palsy. In a study of 28 patients for whom treatment with acyclovir and prednisone was initiated within 3 days after the onset of facial paralysis, the recovery from paralysis was complete in 21 (75%), whereas complete recovery was achieved in only 7 out of 23 patients (30%) for whom treatment was initiated more than 7 days after the onset (30%). The difference betwwen the two groups in terms of facial nerve recovery was significant. The aim of this case report is to emphasize the importance of careful examination and early initiation of therapy in suspected cases of Ramsay Hunt syndrome.

Key Words: Ramsay Hunt syndrome; varicella zoster virus; facial nerve diseases

ÖZET Kraniyal sinir felci, çocukluk yaş grubunda nadir rastlanılan bir bulgudur. Çocuk ve erişkinlerde yapılan tanısal çalışmalar sonucunda varisella zoster virüsü (VZV) reaktivasyonunun 7. kraniyal sinir felcine yol açabileceği kanıtlanmış bulunmaktadır. Virüsün reaktivasyon bölgesi genikülat gangliyondur. Virüs gangliyona büyük olasılıkla suçiçeği sırasında, fasiyal sinirin kulak ve dilde lokalize olmuş sensöriyal dalları aracılığıyla girmektedir. Geriye dönük çalışmalarda, Bell paralizisinin diğer nedenlerine kıyasla, VZV ile ilişkili fasiyal paralizilerin prognozunun daha kötü olduğu gösterilmiştir. Ramsay Hunt sendromu, kulak (zoster otikus) veya ağızda eritematöz vezikülün eşlik ettiği periferik fasiyal sinir felcidir. Bu nedenle VZV reaktivasyonundan şüphe edildiğinde asiklovir tedavisi önerilmektedir. Varisella zoster virüsü reaktivasyonu şüphesinde detaylı kulak muayenesi yapılması büyük önem taşımaktadır. Bu durumda ampirik asiklovir tedavisi başlanabilmektedir.

Anahtar Kelimeler: Ramsay Hunt sendromu; varisella zoster virüsü; fasiyal sinir hastalıkları

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Facial palsy is rare in childhood and varicella infection leading to Ramsay Hunt syndrome may be its cause. Ramsay Hunt syndrome is a lower motor neuron weakness of the seventh (facial) cranial nerve (CN) caused by the reactivation of herpes zoster virus. Around 20-30% of individuals will be affected by herpes zoster during their lives. Ramsay Hunt syndrome is a rare disease, affecting less than 1 in 1500 people in the United States.[1]

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The association of facial palsy with varicella-zoster virus was first described in a 17-month–old child. Ramsay Hunt has studied herpetic inflammation of the geniculate ganglion[2] and described for the first time, the relation between somatic sensory function in the ear and the geniculate ganglion. His discovery of the general sensory function of the facial nerve revealed that functions of the seventh cranial nerve were mixed and included brachial and visceral motor as well as special and general sensory components, one to the pinna for general sensation and another to the tongue for taste sensation. It is presumed that during chickenpox viruses within the vesicles on either the pinna or the tongue might enter the sensory branches of the facial nerve and travel to the geniculate ganglion, where they establish latency. In Ramsay Hunt syndrome, VZV reactivating in the geniculate ganglion and travelling in the sensory branches of CN VII causes inflammation not only in the CN VII motor branches but also in the adjacent acoustic (CN VIII) nerve by a by-stander effect, thus leading to hearing defects. Retrospective reviews suggest that patients with VZV-related facial nerve palsy have poorer outcomes than other cases of Bell's palsy.[3] In this article, we reported two children presenting with facial palsy symptoms and vesicules on the auricula to emphasize the importance of early initiation of acyclovir in achieving promising results.

CASE REPORTS

CASE I

A 12-year-old male patient presented with left ear pain and facial weakness on the left side of the face for the last 7 days. He had had varicella when he was 3 years old. He was prescribed oral acyclovir with a diagnosis of Ramsay Hunt syndrome and his earache had regressed with treatment, but facial weakness had persisted with pain on left side of the face. Laboratory examination was normal and physical examination revealed adherent crusts on the left auricula and left peripheral facial palsy (Figure 1 and 2). He was hospitalized and intravenous acyclovir 1500 mg/m^2, 3 times a day and peroral prednisone 1 mg/kg/day for 5 days (followed by a 10 day taper) was initiated; after 7 days of treatment he recovered completely and was discharged.

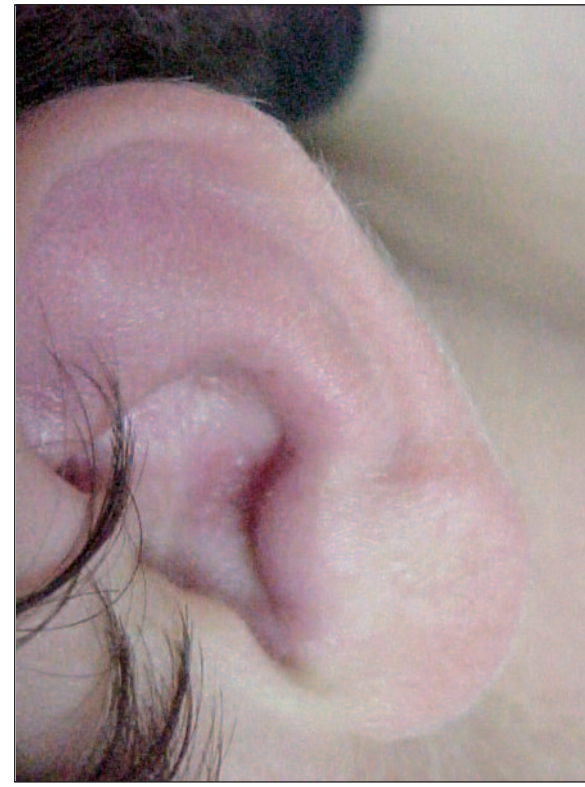

FIGURE 1: Vesicules on the ear.
(See for colored form http://tipbilimleri.turkiyeklinikleri.com/)

CASE II

Our second case was a 10-year-old female patient presenting with facial asymetry and inability to close her left eye. She had had varicella when she was 2 years old. She had tinnitus and itching of the left ear for the last 3 days. Laboratory examination revealed normal findings. Other than vesicular eruption on her left auricula and left peripheral facial palsy, physical examinaton was normal. Minimal sensorineural hearing loss was present on audiologic study. Peroral prednisolone 1 mg/kg/day for 5 days (followed by a 10 day taper) and intravenous acyclovir 1500 mg/m^2, 3 times a day was initiated and she recovered completely on sixth day of admission.

DISCUSSION

Ramsay-Hunt Syndrome (RHS) is a rare form of zoster disease characterized by peripheral facial

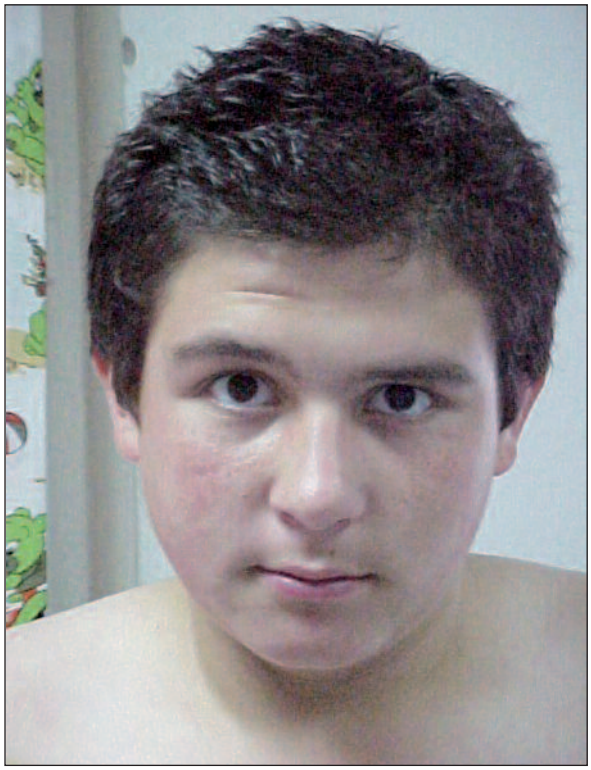

FIGURE 2A: Facial nerve palsy on left side.
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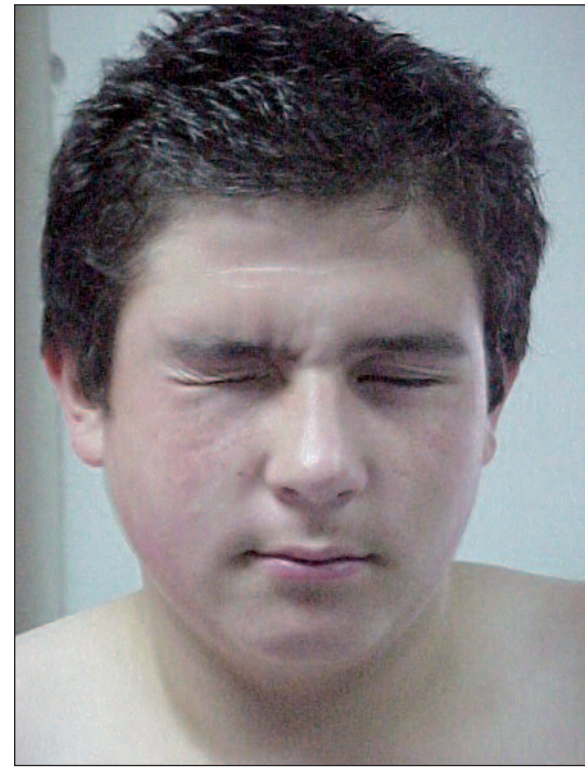

FIGURE 2B: Weakness in closing left eye due to facial nerve palsy on left side.
(See for colored form http://tipbilimleri.turkiyeklinikleri.com/)

weakness, skin eruption in the auricular canal and cochleovestibular symptoms. It should be suspected in patients with facial weakness, erythema, vesicles and/or auricular pain.[4] Strong radiologic evidence supports the concept of VZV latency and reactivation in the geniculate ganglion. Patients with idiopathic (Bell) facial palsy and Ramsay Hunt syndrome have been examined by magnetic resonance imaging after injection with gadolinium.[5] The normal facial nerve shows only minimal enhancement of the geniculate ganglion. In contrast, during Ramsay Hunt syndrome, intense enhancement of the geniculate ganglion is observed. In addition, enhancement of CN VIII may also be noted. Radiologists have speculated that the enhancement is caused by inflammation-induced breakdown of the blood-peripheral nerve barrier and subsequent diffusion of contrast material into the endoneural space.[5] Viral diagnostic analyses clearly indicate that VZV reactivation is an important cause of facial nerve palsy in children, adolescents and adults.[6] In a study to review the initial lesion in

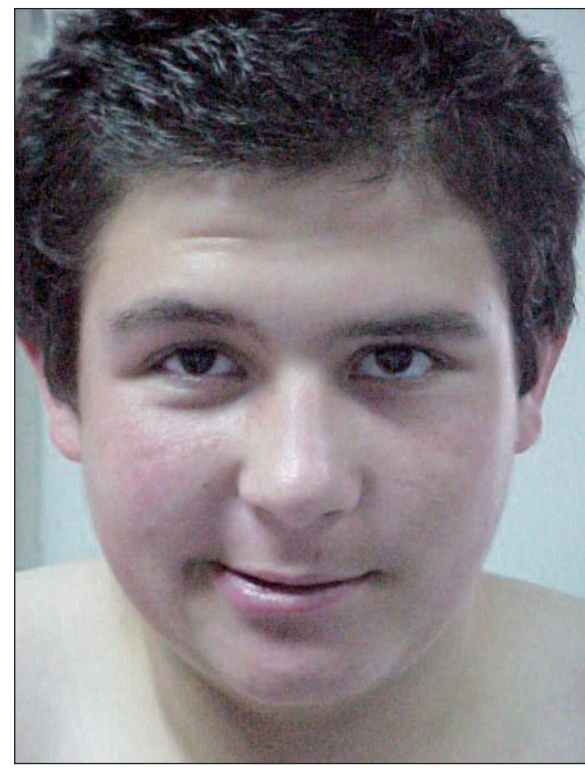

FIGURE 2C: Asymmetry on left side.
(See for colored form http://tipbilimleri.turkiyeklinikleri.com/)

Bell's palsy and Ramsay-Hunt syndrome using intraoperative monitoring of the antidromic facial nerve response (AFNR) revealed that the initial lesion in Ramsay Hunt syndrome was mainly located around the geniculate ganglion within 1 week after the onset of paralysis AFNR.[7]

Because of the clinical impression that VZV-related cases of CN VII palsy have poorer outcomes than all other cases of Bell's palsy, treatment of VZV related facial palsy with oral acyclovir (80 mg/kg/ day) or intravenous acyclovir (45 mg/kg/day) is suggested.[7] Reserving prednisone for the most severe cases and prescription together with acyclovir is recommended.[8] Our two cases were treated immediately with both acyclovir and prednisone. The largest retrospective Ramsay Hunt syndrome treatment study showed a statistically significant improvement in patients treated with prednisone and acyclovir within 3 days of onset.[9] Eighty patients were separated into groups based on the time treatment was started-less than 3 days, 3-7 days, and after 7 days. All patients were treated with oral prednisone (1 mg/kg/day for 5 days followed by a 10 day taper), as well as with intravenous acyclovir (250 mg three times daily), or oral acyclovir (800 mg five times daily). Patients were followed up for 6-12 months with repeated clinical examinations, nerve excitability testing, and audiograms in patients who complained of tinnitus or hearing loss. Complete recovery was achieved in 21 (75%) patients treated within the first 3 days ($p<0.05$), 14 (48%) patients treated at 4–7 days, and seven (30%) cases when treatment was not initiated until after 7 days. Moreover, 26 (50%) patients who were not treated within the first 3 days progressed to a complete loss of response to facial nerve stimulation. No statistically significant differences were noted between patients treated with intravenous or oral acyclovir. Of 12 patients with mild to moderate hearing loss, that were followed up with serial audiograms, six recovered completely, three had partial recovery, and three remained stable. Audiological outcome did not differ significantly between the treatment groups. Despite the lack of randomized controlled prospective treatment trials for Ramsay Hunt syndrome, data from the collective case reports and retrospective reviews suggest that both prednisone and acyclovir, if given early, improve the overall prognosis. The combination of an antiviral with a steroid, with the steroid given after 5 days, was shown to be the most effective pharmacologic management in the study of Coulson et al. Although microvascular decompression and rhizotomy are therapeutic options, they have usually been reserved for resistant neuralgic syndromes.[10,11] Acyclovir was the first medication licensed for the treatment of herpes zoster. Acyclovir therapy accelerated the cessation of new lesion formation and cutaneous healing.[12] Role of electroneurography is controversial; electroneurography performed between day 7 and 10 for Bell's palsy or day 10 and 14 for herpes zoster oticus does not provide accurate information on the prognosis or recovery rate of the facial paralysis.[13]

In a study from Turkey, 15 children with herpes zoster infection were reported; eight cases were treated with parenteral acyclovir, four with oral acyclovir and two with oral valacyclovir. Two patients had Ramsay-Hunt syndrome similar to our cases; none had post-herpetic neuralgia.[14] Facial palsy; which is rare in childhood should remind the likelihood of VZV activation and detailed questioning of earache and ear examination should be done immediately. Although the treatment of choice in Bell's palsy is steroid, since accurate data about steroid use in Ramsay Hunt syndrome is lacking and may cause aggravation of viral spreading, acyclovir treatment should be initiated in the early period.

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