Case Report

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Ramsay Hunt Syndrome in a Living-donor Kidney Transplantation Recipient: Unusual Clinical Course Case

Dawn Jung, M.D.[1], Sung Hoon Kim, M.D.[1], Seung Ok Choi, M.D.[2] and Sang You Park, M.D.[3]

Departments of Surgery[1], Internal Medicine[2] and Otorhinolaryngology[3], Wonju Severance Christian Hospital, Yonsei University Wonju College of Medicine, Wonju, Korea

Varicella zoster virus (VZV) infection is due to VZV reactivation in most cases. The infection rate ranges from 4% to 12% in renal allograft recipients. Ramsay Hunt syndrome (RHS) is a rare manifestation of VZV infection. RHS typically presents as severe ear pain, small vesicles, and facial palsy. We reported a case of a 60-year-old man with an unusual clinical course who underwent living donor renal transplantation. He complained of severe ear pain but did not show vesicles or facial palsy. He also presented lesions indicating a fungal infection. Diagnosis of RHS was delayed since facial palsy did not develop until some days later. Although the denervation rate was high, he showed recovery of nearly all symptoms after antiviral treatment. Solid organ recipients may not typically show presentation of viral infection, and therefore clinical suspicion is important. Even though the final diagnosis is delayed, we must treat patients since they may recover well in contrast with the average population.

Key Words: Kidney transplantation, Human herpesvirus 3, Herpes zoster oticus
중심 단어: 신장이식, 제3형 사람 헤르페스 바이러스, 귀대상포진

INTRODUCTION

Varicella zoster virus (VZV) has two distinct infection patterns in solid organ transplantation recipients(1). VZV reactivation is the most common pattern and causes herpes zoster in seropositive recipients. The incidence of VZV infection ranges from 4% to 12% in renal allograft recipients(2). It usually causes vesicular lesions and pain in a dermatome. If VZV in geniculate ganglion is reactivated and affects the seventh and eighth cranial nerves, it causes herpes zoster oticus and acute peripheral facial paralysis. It is known as Ramsay Hunt syndrome (RHS)(3). Clinical presentations of RHS are severe ear pain, small vesicle, and facial palsy in immune-compromised patients. However, RHS is an unusual disease entity in solid organ transplantation recipients. There is only one case report about RHS in Korea(4). That patients showed typical symptoms including ear pain and vesicle and was easily diagnosed as having RHS. We have experienced patients who showed atypical symptoms such as general weakness, easy fatigue, and ear pain and the diagnosis of RHS was delayed. However, we successfully treated the patient with unusual course RHS although the diagnosis of RHS was delayed and the denervation rate was so high. We present the patient with unusual course RHS.

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Corresponding author: Sung Hoon Kim
Department of Surgery, Wonju Severance Christian Hospital, Yonsei University Wonju College of Medicine, 20 Ilsan-ro, Wonju 26426, Korea
Tel: 82-33-741-0573, Fax: 82-33-741-0574
E-mail: drgs01@yonsei.ac.kr

CASE REPORT

A 60-year-old male underwent kidney transplantation in 2012 due to polycystic kidney disease from a living unrelated donor (his spouse). Preoperative viral serologic test results showed VZV immunoglobulin (Ig) G was positive

and IgM was negative. After kidney transplantation, he was discharged at postoperative 9 days with normal kidney function and without any rejection or infection episodes. Basiliximab was used as induction therapy on the operation day and postoperative day 4. His immunosuppression regimen included tacrolimus (1 mg twice daily), prednisolone (5 mg twice daily), and mycophenolate sodium (720 mg twice daily). One year after transplantation, he visited an outpatient clinic and complained of general weakness, easy fatigue, and ear pain for the previous 5 days. Laboratory data showed a white blood cell count of 2,750 cells/mm^3, tacrolimus trough level 5.0 ng/mL, and serum creatinine 0.94 mg/dL. His tongue was covered by a white patch and there was a small amount of serous discharge from the left pinna without vesicle. An otorhinolaryngologist suspected an anti-fungal infection and performed a fungal smear, but the result was negative. We stopped the prescription of mycophenolate sodium and prophylactically prescribed an anti-fungal agent. Five days later, he complained of more severe symptoms and also dizziness. Brain computer tomography and magnetic resolution images did not show any abnormal findings. Three days later after admission, facial palsy developed and then we diagnosed RHS with identification of elevated VZV IgM titer. The VZV IgM titer was changed from preoperative 0.3 to 4.4. Electroneurography showed involvement of left facial nerve with 95.65% denervation ratio. The otorhinolaryngologist prescribed acyclovir and methylprednisolone. Acyclovir was administrated at a dose of 250 mg intravenous five times daily for 7 days. Methylprednisolone was given as 48 mg through mouth for 7 days and tapered and stopped as half dose at a 3 days interval. And acyclovir was given as 800 mg through mouth five times daily for 21 days. After 4 weeks treatment, he showed near complete resolution of facial palsy and other symptoms. Mycophenolate mofetil was restarted 2 months after its initial stop. His creatinine level was maintained below 0.9 mg/dL during all treatment period and continues to be.

DISCUSSION

VZV infection has two distinct courses after transplantation. One is primary infection in seronegative patients and the other is reactivation in seropositive patients. Nearly ninety percent of solid organ transplant patients show VZV reactivation. The incidence of VZV infection in kidney transplant patients ranged from 4% to 12%(2). Herpes zoster oticus with acute peripheral paralysis is known as RHS in 1907(3). RHS is caused by the reactivation of VZV in the geniculate ganglion and represented as the dysfunction of the facial and vestibulocochlear nerve(5). RHS is not a rare disease and shows incidence in about five cases per 100,000 in the United States population. However, RHS is very rare disease in solid organ transplant patients and as far as I know, only seven RHS cases in renal allograft patients were reported(4,6-11). The incidence of RHS increased in patients older than 60 years(12). However, all kidney transplant patients except our patient and one another patient, showed young age patients as less than 40 years. Clinical suspicion is most important because diagnosis is based on clinical signs and symptoms. Typical vesicle and facial palsy are diagnostic clues of RHS. Early treatment using anti-viral agents and steroids may reduce sequela of RHS. The initiation of antiviral therapy is most effective within the first 72 hours of symptom onset and the recovery rate of facial palsy was reported as 75%(3). The diagnosis of RHS was delayed in many kidney transplant patients because of unusual presentation. However, most studies reported a nearly resolved condition of facial palsy unlike the results of the general population. Higher denervation rate may show higher sequela rate(13).

In our case, diagnosis of RHS was delayed due to late development of facial palsy and abscess of vesicle. Even though anti-viral and steroids were started 13 days later after symptom development and a high denervation rate was shown, he successfully recovered. Solid organ recipients may not typically show presentation of viral infection and therefore clinical suspicion is important. Even though the final diagnosis is delayed, we must treat the patients actively because the patients may recover well, in contrast with the general population.

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