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Coexistence of Atypical Ramsay Hunt Syndrome and Varicella-Zoster Virus Encephalitis

Atipik Ramsay Hunt Sendromu ve Varisella-Zoster Virüs Ensefaliti Birlikteliği

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Dear Editor,

Varicella-zoster virus (VZV) is a DNA virus transmitted via droplets. It lies latent in neurons and causes zona zoster, central nervous system infections or Ramsey Hunt syndrome (RHS) by reactivation (1). Classic RHS is characterized by herpes zoster oticus and unilateral facial paralysis (2,3,4). Rarely, V., VI., IX., X., XI. and XIIth cranial nerve palsies have been reported in the literature (1).

VZV encephalitis is characterized by impairment of consciousness, focal central nervous system symptoms, and clinical and laboratory findings of herpes zoster [positive cerebrospinal fluid (CSF) VZV DNA and anti-VZV IgM results or four times increase in anti-VZV immunoglobulin-G titers measured two weeks apart] (2). Incidences of RHS and VZV are both low and their coexistence is rarely observed (2). In this article, a patient who presented with multiple cranial nerve palsies and was diagnosed with RHS and VZV encephalitis is discussed.

A 34-year-old Afghan male admitted with new-onset diplopia who had pain in his left ear and throat, tinnitus, vertigo, and facial weakness, which he had had for three weeks. The first neurologic examination showed limitation of outward movements of his right eye, right peripheral facial paralysis, and hypoesthesia of the right side of his face. Dysphagia, imbalance, and hiccups were added in the follow-up, and a neurologic examination on the ninth day showed limitation of downward movements in his right eye and vertical nystagmus, mild deviation of his tongue to the right, paresis of the right palatal arch, and left Babinski sign. The patient had left focal-onset secondary generalized seizures on the tenth day. Meanwhile, herpetic vesicles appeared on both his auriculas and inside both meatus acusticus externus (Figure 1).

Contrast-enhanced cranial magnetic resonance imaging (MRI) at admission was normal. Cranial MRI was repeated after left focal-onset secondary generalized seizures and non-enhancing lesions in the right temporal and insular region, which were suggestive of encephalitis (Figure 2, 3, 4). Electroencephalography showed

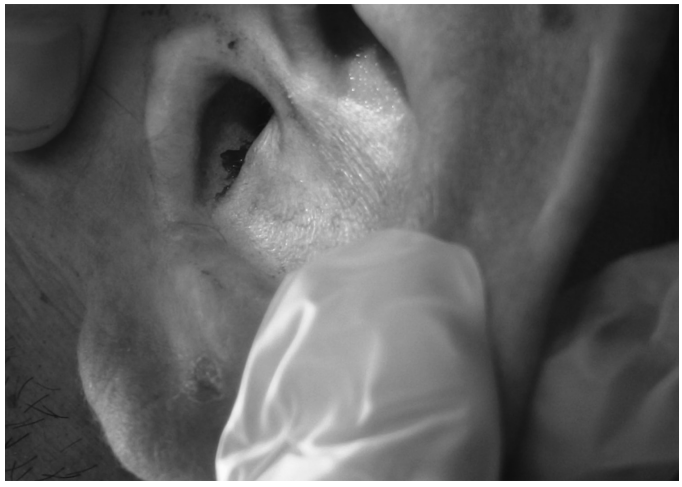

Figure 1. Hemorrhagic vesicles on the auricula and in the meatus acusticus externus

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right temporal focal-onset secondary generalized epileptiform abnormality. No cells were seen in the CSF, and biochemical tests of the CSF were normal.

Progressive cranial nerve involvements, seizures, fever, and lesions in the right temporal and insular cortices in the cranial MRI suggested encephalitis caused by atypical RHS. Toxoplasmosis, rubella cytomegalovirus, herpes simplex, and HIV (TORCH) panels revealed herpes simplex with positive VZV IgM and IgG serology. A search for immunosuppression, tuberculosis, sarcoidosis, and vasculitis was negative.

Valacyclovir 300 mg/day for five days was administered. Additionally, prednisolone 80 mg/day and levetiracetam 1000 mg/day were administered. The patient's clinical and neurologic symptoms improved in three weeks after the treatment and therefor steroid therapy was tapered.

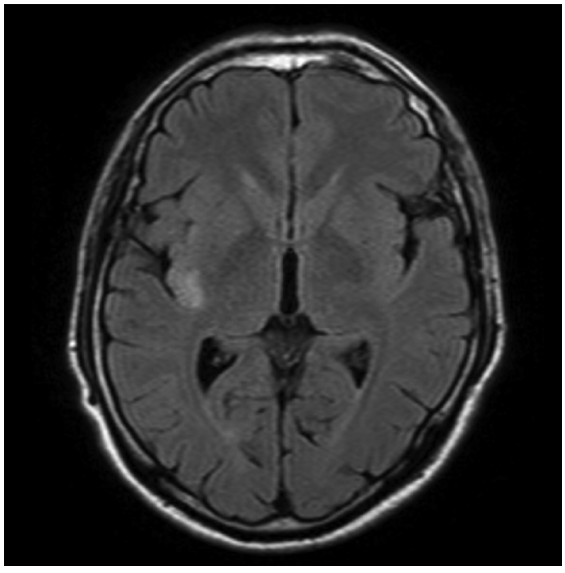

Figure 2. Hyperintense lesion in the right temporal and insular region in axial FLAIR-weighted magnetic resonance images

Erythematous ear lesions and unilateral peripheral facial nerve palsy are seen in typical RHS and VIIIth cranial nerve impairment frequently accompanies (1) the clinical picture. V., VI., IX., X., XI., and XIIth cranial nerve impairments and also trochlear nerve impairment can be seen very rarely in atypical cases (1,5).

Direct perineural spread of VZV via anastomotic pathways or vasculitic involvement can cause multiple cranial nerve palsies during VZV reactivation (3). Inflammatory involvement can be shown by neuro-imaging in some patients (1,3).

Glossopharyngeal/vagal symptoms are rarely seen in RHS. Although the pathogenesis is not well known, it is suggested that VZV may reach nerve terminals by crossing the mucosa and indirectly infecting the IXth and Xth cranial nerves following a laryngopharyngeal infection or VZV might spread via nerve anastomoses (1). Although we observed no radiologic findings, we think that dysphagia may have been caused by IXth and X cranial neuropathies in our patient.

Co-existence of RHS and VZV encephalitis is not reported frequently in Western countries but is reported more in Japan, which suggests that racial differences may play a role (2). The Afghan origin of our patient may play a role in the co-existence of RHS and VZV encephalitis.

Multiple cranial nerve involvement can be seen in atypical RHS, and herpetic lesions, which are essential for the diagnosis of RHS, can appear after cranial neuropathy. Similarly, radiologic findings cannot be detected in the early period. We report this patient to highlight that multiple cranial nerve involvement can accompany RHS, and that RHS and encephalitis can coexist.

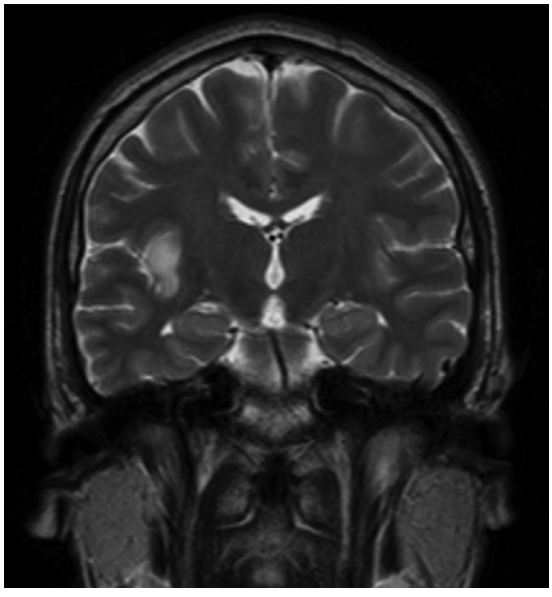

Figure 3. Hyperintense lesion in the right temporal and insular region in coronal T2-weighted magnetic resonance images

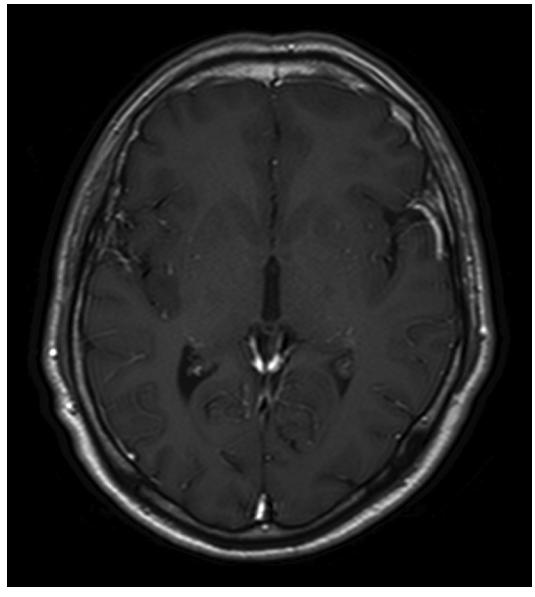

Figure 4. No contrast enhancement in axial T1-weighted magnetic resonance images with contrast

Ethics

Informed Consent: Consent form was filled out by all participants.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Concept: B.K., S.S.Ç., Design: B.K., Data Collection or Processing: B.K., A.S.E., Analysis or Interpretation: B.K., S.S.Ç., Literature Search: B.K., Writing: B.K., A.S.E.

Conflict of Interest: No conflict of interest was declared by the authors.

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