Dear Sir,

A 55-year-old gentleman working as a farmer presented to the emergency department with sudden onset of left ear pain with erythematous rashes, which were initially vesicular and then gradually scabbed. On the 3rd day of rash, the patient developed bilateral facial paresis, flaccid dysarthria, dysphagia, and on the following day, he developed gait ataxia. He denied fever, headache, nuchal rigidity, altered consciousness level, vertigo, weakness, or other neurological complaints. He was not diabetic or hypertensive. He was a nonalcoholic and a nonsmoker.

Plain computerized tomography (CT brain) done at another center was interpreted as normal. His cerebrospinal fluid analysis showed six cells with normal proteins and glucose. Qualitative polymerase chain reaction (PCR) for varicella zoster virus (VZV) in cerebrospinal fluid (CSF) was positive. Magnetic resonance imaging (MRI) showed T2 and fluid attenuation inversion recovery (FLAIR) hyperintense signal in the left dorsal medulla and pons [Figure 1], traceable to the intraparenchymal course of the left facial nerve, which was mildly enhancing on postgadolinium contrast. Susceptibility-weighted imaging (SWI) images did not reveal

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**Ramsay Hunt Syndrome Leading to the Brainstem and Cerebellar Involvement: A Case Report of a Rare Co-occurrence**

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any blooming. Axial diffusion-weighted images revealed no restriction in the lesion. The angiogram was normal.

He was initiated on intravenous methylprednisolone 1 g/day for 5 days and intravenous acyclovir at 30 mg/kg/day in three divided doses for 7 days followed by tablet valacyclovir 1 g twice daily for 7 days. Five days after the treatment, his gait ataxia improved with mild improvement in speech.

His routine blood parameters, including complete blood count, renal function tests, liver function tests, sodium, and potassium, were normal. Serum B12 levels, folate levels, and homocysteine levels were normal. His human immunodeficiency virus (HIV) by enzyme-linked immunosorbent assay (ELISA), hepatitis B surface antigen (HBsAg), hepatitis C virus (HCV), and venereal disease research laboratory (VDRL) were nonreactive, and glycated hemoglobin (HBA1C) was 4.9%. Anti-nuclear antibody (ANA) and anti-neutrophil cytoplasmic antibody (ANCA) profiles were negative. The absolute CD4 count was 52 cells/µL. Pure tone audiometry showed a mild sensorineural hearing loss on the left side.

**Discussion**

James Ramsay Hunt had first described this eponymous syndrome in 1907. Ramsay Hunt had hypothesized that Gasserian, geniculate, petrous, accessory, jugular, plexiform, and...
and second and third cervical dorsal root ganglia comprised a chain of contiguous ganglia.\textsuperscript{[1]} This could explain the associated tinnitus, hearing loss, nausea, vomiting, vertigo, and nystagmus found in some cases. A retrospective study of 102 patients with Ramsay Hunt Syndrome (RHS) done in 1988 showed that only 10% of the patients achieved complete recovery.\textsuperscript{[2]} Varicella-Zoster vasculopathy (VZV) is a distinct complication of zoster infection leading to infarction or hemorrhage with MR angiogram abnormalities. Peitersen had reported 2,500 cases of peripheral facial nerve palsies, and in this study, a complete recovery was achieved only in 21% of the herpes zoster oticus cases.\textsuperscript{[3]} Predisposing factors for reactivation include diabetes mellitus, spinal anesthesia, malignancies, and conditions associated with immune suppression such as lymphoma, steroid therapy, immunosuppressive agents, and acquired immunodeficiency syndrome (AIDS). However, none of these were evident in our patient. We describe an immunocompromised patient who developed brainstem and cerebellar involvement following RHS. Our patient presented with vesicular rash and left ear pain, following which he developed bilateral facial palsy, flaccid dysarthria, and dysphagia, which suggested brainstem involvement. He then developed gait ataxia, which suggested cerebellar involvement. Acute cerebellar involvement following VZV infection is well known in children, however, rare in adults. The MRI features, CSF analysis, and pure tone audiometry also correlated with the clinical picture. After 5 days of intravenous pulse methylprednisolone and intravenous acyclovir, the patient showed significant improvement in gait ataxia and mild improvement in dysphagia. Timely initiation of steroids and antiviral agents is the key to recovery.\textsuperscript{[4]} However, dysarthria and bilateral facial paresis did not improve. The prognosis of facial palsy in RHS remains poor.\textsuperscript{[5]} The recovery in this patient can be attributed to the pharmacological intervention or the natural course of the disease. The neurological illness being due to axonal spread of the virus, rather than a vasculopathy is supported by the fact that the course of the disease was progressive, rather than multiple transient ischemic attacks, with no evidence of diffusion restriction or angiogram abnormality on magnetic resonance imaging, and the T2 hyperintensities were not respecting strict vascular territories. A similar observation was noted by Hu \textit{et al.} in 2003.\textsuperscript{[6]} The CSF VZV PCR is sensitive as well as a specific biomarker for zoster infection with neurological symptoms.\textsuperscript{[7]} It has been postulated that mutations in genes participating in the toll-like receptor 3 (TLR3) pathway may predispose to encephalitis caused by VZV.\textsuperscript{[8]}

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**Conflicts of interest**
There are no conflicts of interest.

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