Ramsay Hunt Syndrome: Clinical Presentation, Diagnostic Challenges, and Treatment Efficacy

Síndrome de Ramsay Hunt: Apresentação Clínica, Desafios Diagnósticos e Eficácia do Tratamento

Keywords: Herpes Zoster Oticus/diagnostic imaging; Magnetic Resonance Imaging; Tomography, X-Ray Computed
Palavras-chave: Herpes Zóster Ótico/diagnóstico por imagem; Ressonância Magnética; Tomografia Computorizada

Dear Editor,

Ramsay Hunt syndrome (RHS), first described by James Ramsay Hunt in 1907, is a rare condition caused by the varicella zoster virus,[1,2] affecting approximately five in 100 000 individuals, typically aged 20 - 30, with no sex differences.[3] It involves the facial and vestibulocochlear nerves due to their proximity, leading to symptoms such as facial paralysis, ear pain, herpes vesicles, and occasionally vertigo, nausea, or nystagmus. Compared to Bell's palsy, RHS often presents with more severe paralysis and delayed rash onset in 14% of cases, complicating diagnosis.[1]

We present the case of a 73-year-old man with a 15-day history of right-sided facial muscle weakness, intense right ear pain, and vesicular eruptions in the external auditory canal and auricle. He had a medical history of diabetes *mellitus*. Physical examination revealed loss of right frontal sulci, difficulty closing the right eye, and mouth deviation to the left. A computed tomography scan revealed an inflammatory process in the tympanic membrane on the right side. A magnetic resonance imaging (MRI) scan detected an inflammatory neuropathy of the right VII cranial nerve (facial nerve) within the internal auditory canal, along with right sided otomastoidopathy and inflammatory changes in the external auditory canal and auricle on the right. Polymerase chain reaction (PCR) detected the varicella zoster virus, confirming RHS. The patient was treated with oral prednisone (1 mg/kg/day for five days, tapered over 10 days), oral acyclovir (800 mg five times daily for seven days), and facial rehabilitation. He showed improvement within 10 days and was symptom-free six months later.

The histopathology of RHS varies with its progression. Post-mortem studies show necrosis of the dorsal ganglion branches in acute cases, while chronic cases reveal fibrosis in the vestibular ganglia.[1] A diagnosis of Ramsay Hunt syndrome relies on clinical history, physical examination, and supportive imaging.[1] While MRI can identify nerve inflammation,[4,5] PCR offers higher sensitivity when using mononuclear blood cells, as demonstrated in the presented case, or samples from the geniculate area.[1] Short-term complications of RHS include corneal abrasions, exposure keratopathy, depression, social anxiety, and the potential spread of chickenpox to unvaccinated or immunocompromised individuals. Long-term complications, such as synkinesis (involuntary contractions), postherpetic neuralgia, vesicular scarring, and persistent psychological effects, are possible but were absent in this case. Treatment typically involves antiviral therapy, with acyclovir being the primary agent. Combining antivirals with steroids has shown better outcomes in improving facial control (70.5%) compared to antiviral monotherapy (68%).[3]

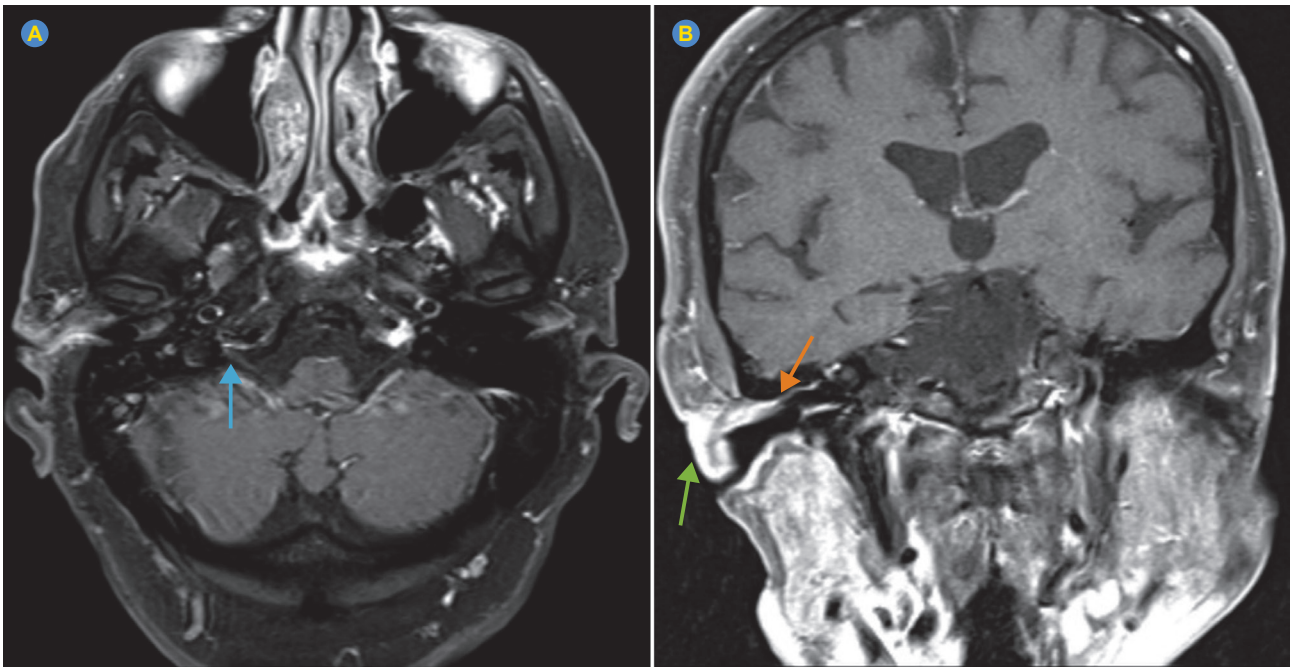

Figure 1 – Magnetic resonance imaging T1 post-contrast sequence in axial section (A) and coronal section (B) demonstrating focal enhancement of the canalicular right facial-vestibulocochlear nerve complex (blue arrow) and an avid enhancement of the right external auditory canal (orange arrow) and external ear/ pinna (green arrow)

AUTHOR CONTRIBUTIONS

All authors contributed equally to this manuscript and approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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