Journal of Medical Research and Health Sciences
Received 29 Jan 2022 | Revised 20 Feb 2022 | Accepted 29 Mar 2022 | Published Online 17 Apr 2022

DOI: https://doi.org/10.52845/JMRHS/2022-5-4-7

JMRHS 5 (4), 1899–1905 (2022)

ISSN (O) 2589-9031 | (P) 2589-9023

Research Article **Open Access Journal**

Typical Mri Findings of Ramsay Hunt Syndrome

Andrés Felipe Díaz Muñoz[1*], Talel Muhieddine Ibrahim[2], Chiara Theresa Norato Ortiz[3], Ángel Fabián Leon Chávez[4], Geinni Paola Barrera Amaya[5], Maria Carolina Causil Galvis[6], Marlon Eduardo Freite Pacheco[7], Miguel Angel Iglesias Herrera[8]

Corresponding Author: Andrés Felipe Díaz Muñoz

[1]4th Year Radiology Resident, Pontifícia Universidade Católica do Rio de Janeiro
[2]General Physician, Universidad Cooperativa de Colombia sede Santa Marta
[3]General Physician, Fundación Universitaria Sanitas
[4]General Physician, Universida del Bosque, Bogotá
[5]General Physician, Universidad del Santander UDES
[6]General Physician, Universidaddel Sinú, Montería
[7]General Physician, Universidad del Norte, Barranquilla
[8]General Physician, Universidad del Sinú, Cartagena

Abstract:
Ramsay Hunt syndrome (SRH) is characterized by its appearance in the reactivation of the Varicella Zoster virus (VZV) infection causing affections in the geniculate ganglion, more specifically in the VII pair, the prevalent age is over 60 years, causing edema, nerve compression and demyelination. This syndrome is generally diagnosed with the patient's clinic where the present history and physical examination play an important role, however, imaging studies are also used mainly in atypical forms, such as contrast-enhanced magnetic resonance imaging (MRI) of the head and neck, which allows a better visualization of compromised structures such as the brain, the base of the skull, the thyroid region and a better contrast uptake of the facial nerve and the geniculate ganglion, thus allowing the assessment of the level of compromise and existing condition

Keywords: Ramsay Hunt Syndrome, Magnetic Resonance Imaging, Varicella Zoster Virus, Findings.

Introduction

Ramsay Hunt syndrome (RHS) was first described at the beginning of the 20th century by James Ramsay Hunt, reporting several cases of patients with erythematous vesicles in the auricular area, in the oral mucosa, and also different neurological alterations. This syndrome is characterized by its appearance in the reactivation of the Varicella Zoster virus (VZV) infection, causing affections in the geniculate ganglion, more specifically in the VII alpha cranial nerve. Herpesvirus is similar to herpes simplex virus type 1 and type 2 with different characteristics. present in its clinical behavior and latency mechanism, humans are the only reservoir of this virus and it is spread directly by contact with VZV skin lesions or by inhalation of respiratory droplets through coughing from sick patients to patients healthy, in 1907 James Ramsay Hunt described three neurological syndromes caused by VZV infection, Ramsay Hunt Syndrome type 1, a very rare cerebellar syndrome that causes cerebellar degeneration and leads to ataxia, myoclonus, tremors and seizures. Ramsay Hunt syndrome type 2, which is the most common type, involves reactivation of the varicella-zoster virus in the geniculate ganglion, leading to vesicular disease, radiculopathy, and ganglitis. There is also Ramsay Hunt syndrome

type 3 (Hunter disease), which is a neuropathy of the ulnar nerve (1,2). This syndrome is produced after alterations in the host's immune system accompanied by risk factors such as an underlying medical condition or age, being more common in those over 60 years of age and rare in those under 6 years of age, causing edema, compression in the nerve and demyelination which increases nerve damage. SRH is considered the second cause of atraumatic facial paralysis with lower motor neuron-type characteristics and 10-12% of cases of acute facial paralysis are attributed to this etiology. The clinical manifestations of this syndrome are linked to the neuronal structure where the reactivation of VZV, however, a classic triad is described that includes: ipsilateral facial paralysis, otalgia, vesicular lesions in the external auricular region and the pinna (3,4).

Taking into account that this syndrome, also called Herpes Zoster oticus, has a variable clinical presentation, which includes both neurological and dermatological symptoms, it has had to be classified into stages, which consists of:

Stage I: Presence of otalgia and vesicular eruption in the auricle.

Stage II: Includes the manifestations of stage I plus ipsilateral peripheral facial paralysis.

Stage III: Includes characteristics of stages I and II plus vestibular symptoms such as tinnitus, hearing loss or vertigo.

Stage IV: Includes the characteristics of the 3 previous stages plus the presence of compromise and alteration of other cranial nerves, mainly the V pair.

The percentage of affection in other cranial nerves is the following V pair (sensory branch) 36%. V pair (motor branch) 11%, VII pair (vestibular branch) 26%, VII pair (cochlear branch) 26%, IX pair (sensory branch) 23%, X pair (recurrent laryngeal) 19% (1,4)

This syndrome is generally diagnosed with the patient's clinic where the present history and physical examination play an important role, however, imaging studies are also used mainly in atypical forms, such as contrast-enhanced magnetic resonance imaging (MRI) of the head and neck, which allows a better visualization of compromised structures such as the brain, the base of the skull, the thyroid region and a better contrast uptake of the facial nerve and the geniculate ganglion, thus allowing to evaluate the level of compromise and existing affection where the findings Mainly found are the correlation between the intensity of contrast uptake in contrast-enhanced NMR images in T1 and the characteristic inflammation present in the facial nerve in the labyrinthine segment, the geniculate ganglion and the pyramidal segment present in patients with SRH, it is also found present enhancement in the facial nerve and intense enhancement in the tympanic and mastoid segment of the facial nerve, related to alterations in the different segments of the nerves due to the degeneration and regeneralization process given by the inflammatory paralysis, these alterations can be present for months and even in patients with clinical recovery it can present The link in the intrameatal distal segment persists, the treatment of this entity is based on symptomatic management with various drug groups including corticosteroids, antivirals, analgesics and the prevention of complications such as postherpetic neuralgia, neuropathies, ophthalmic herpes zoster encephalitis, cerebellitis and myelitis, thus considering that a timely diagnosis of this syndrome contributes to providing adequate and timely care that prevents future complications (5,6).

Materials and methods:

To carry out this article, a bibliographic search was carried out in various databases such as Elsevier, Scielo, Medline, pubmed, ScienceDirect and Ovid, thus selecting original articles, case reports and bibliographic reviews from 2009 to 2021 in Spanish and English using MeSH terms: exercise, cardiovascular dynamics, strength, and using the Boolean operators: and and or. Thus including all the documents that will deal with the impact of strength exercises on cardiovascular dynamics and information related to it, the data found were between 8-14 records, thus using 10 articles for the preparation of this document.

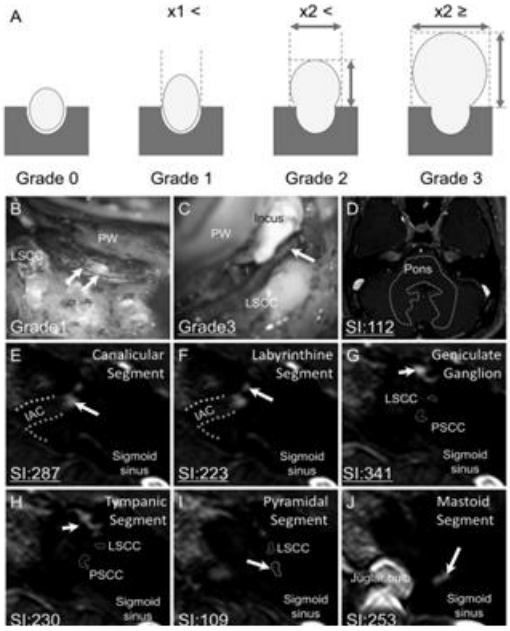
Results:

Based on studies, it has been shown that contrast-enhanced nuclear magnetic resonance imaging of the head and neck is very useful in the evaluation of the brain, skull base, petrous bone, and parathyroid region, as well as to obtain a better visualization of the uptake of contrast by the facial nerve, mainly at the level of the facial foramen, the labyrinthine portion and the geniculate

ganglion, in order to observe the level of affectation correlated with contrast uptake (5).

In a study conducted on 16 patients at the University of Nagoya Japan in 2018, they found a positive correlation between the intensity of contrast uptake in contrast-enhanced MR images in T1 and inflammation of the facial nerve in the labyrinthine segment, the geniculate ganglion and the pyramidal segment in patients with Ramsay Hunt syndrome. They concluded that contrast-enhanced MRI is a useful study to assess the severity of facial nerve involvement by the varicella zoster virus, as well as to predict patient recovery (7).

Figure 1. Evaluation of the severity of facial nerve inflammation in the region of interest in each segment of the facial nerve in magnetic resonance images. (A) System used to measure the degree of inflammation of the facial nerve intraoperatively. Grade 0, no swelling; grade 1, the diameter of the inflamed nerve does not exceed that of the original facial canal; grade 2, the diameter of the inflamed nerve exceeds that of the original facial canal but not by twice; grade 3, the diameter of the inflamed nerve exceeds twice the diameter of the original facial canal. (B, C) Representative images of the inflamed nerve during facial nerve decompression surgery in patients with Ramsay Hunt syndrome. Images were obtained after opening the epineurium of the facial nerve. (B) A mastoid segment with grade 1 nerve inflammation. (C) A pyramidal segment with grade 2 nerve inflammation. LSCC, lateral semicircular canal; PW, posterior wall. (D–J) Representative images of the signal intensity of the central area of the cerebellum on the same image, SIROI (non-CE) is the average SI of the ROI at the equivalent location on the non-contrast-enhanced T1-weighted image, and SIcerebellum (non CE) is the average SI of the central area of the cerebellum in the same image. The SI of the normal facial nerve is enhanced by contrast. Improves to varying degrees depending on the segment (7).



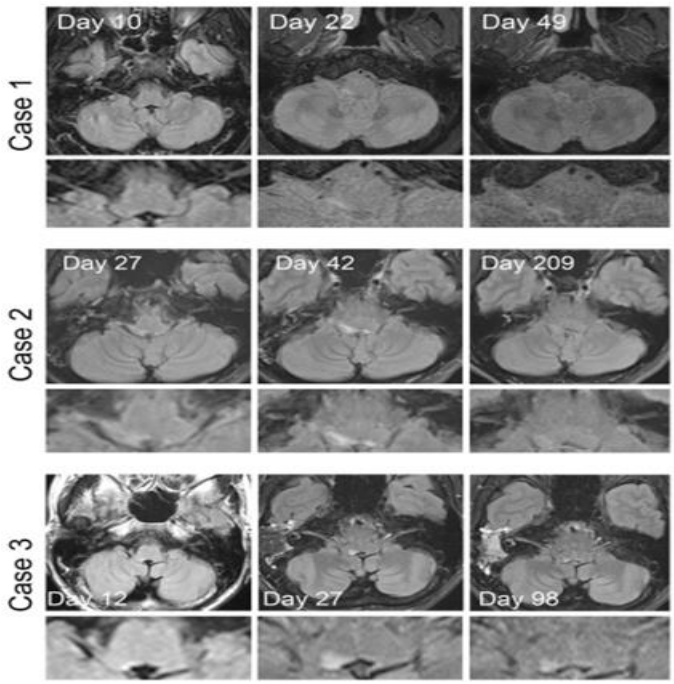
Taken from: Contrast enhanced magnetic resonance imaging of facial nerve swelling in patients with severe Ramsay Hunt syndrome.

In a study carried out in 2008, Adachi described the magnetic resonance findings of nerve IX and X, with thickening and enhancement in varicella zoster virus infection. MRI findings included thickening and contrast enhancement of the IX and X cranial nerves in 2 patients, contrast enhancement of the V nerve in 1 patient, lymph node enhancement in 2 patients, and enhancement of the anterior condylar canal in 1 patient. Inflammation of cranial nerves IX and X can be caused by edema and inflammatory cells, such as previously reported inflammatory infiltrates of lymphocytes and plasma cells in the geniculate ganglion of the facial nerve (8).

In a case report from 2018 in Japan, three cases of Ramsay Hunt syndrome manifesting with brainstem lesions on FLAIR images were presented. The first case initially showed no abnormalities on MRI and cases 2 and 3 showed weak signals on the first MRI that subsequently reached a plateau. These observations suggest the

time frame within which it is possible to detect regional and temporal evolution, that is, that the distribution of affected regions expands between weeks 2 and 5 after the onset of paralysis. All regions of the lesions represent specific neuronal structures: the ipsilateral solitary nucleus (SN), the spinal trigeminal nucleus and tract (STNT), and the vestibular nucleus. The distribution can reasonably be explained by transsynaptic spread of varicella zoster virus disseminated from cell bodies in the geniculate ganglion, from where the two primary lesions receive afferent fibers, including the contralateral lesion in the caudal end of the NS that receives afferent input from the contralateral geniculate ganglion. . In addition, the vestibular nucleus receives input from the vestibular primary afferent ganglia, where latent varicella zoster virus is found in some patients (13).

Figure 2. Progression of brainstem lesions in Ramsay Hunt syndrome on FLAIR images at the same level as the rostral spinal cord.



Taken from: Ramsay Hunt syndrome associated with solitary nucleus, spinal trigeminal nucleus and tract, and vestibular nucleus involvement on sequential magnetic resonance imaging.

Among the 19 cases with brainstem involvement reported so far (Clerk-Lamalice et al. 2015; Ricigliano et al. 2017), 8 had positive MRI findings; 4 of these 8 cases had similar patterns in the affected regions, namely the SN, STNT and vestibular nucleus. These observations indicate that these regions are relatively prone to developing encephalitis after HSR (13).

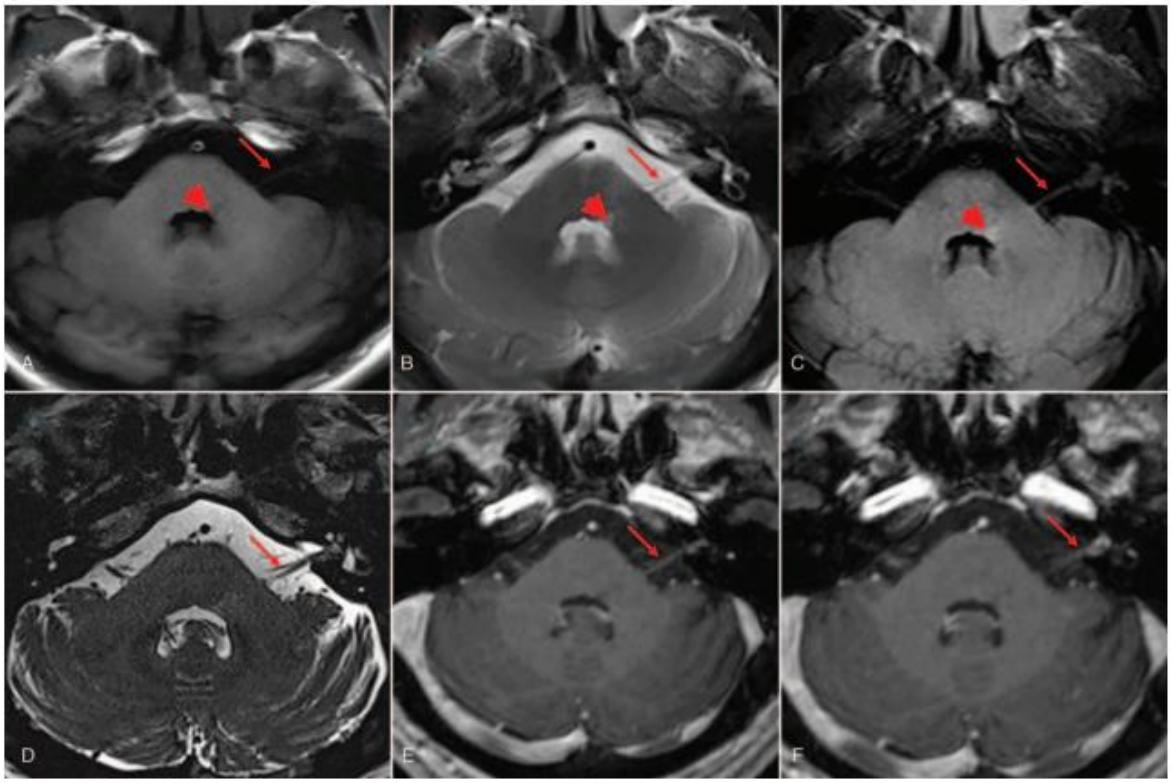
Table 1. Published cases describing abnormal brainstem MRI findings in Ramsay Hunt syndrome.

Article		First detection on MRI (days after onset of facial paralysis)	Lesions on FLAIR			
			Solitary nucleus	Spinal trigeminal nucleus and tract	Vestibular nucleus	Other areas
Mizock BA et al. 2000		8				Pons
Nogueira RG et al. 2003		44		+		
Hu S et al. 2004		3				Superior cerebellar peduncles
Kim JH et al. 2007		31	+		+	Ambiguous n.
Kasai D et al. 2012		n/a	+		+	
Calabria F et al. 2014		~9				Pons, midbrain, and periventricular area
Clerk-Lamalice O et al. 2015		−4 (Before onset)		+		
Shen YY et al. 2015		4				Basis pontis, medulla oblongata
Our cases	Case 1	22	+	+		
	Case 2	27	+ (Bilateral)	+	+	
	Case 3	12	+		+	

Taken from: Ramsay Hunt syndrome associated with solitary nucleus, spinal trigeminal nucleus and tract, and vestibular nucleus involvement on sequential magnetic resonance imaging

In a 2019 case report, post-admission brain MRI findings revealed long T1 and T2 signals, high FLAIR, and diffusion-weighted imaging (DWI) signals in the left peduncle cerebellaris medius. The DWI revealed a high signal. Inflammation of the facial nerve and the vestibulocochlear nerve. The DWI revealed a high signal along the nerve pathway. The gadolinium-enhanced MRI revealed that the facial nerve and the vestibulocochlear nerve at the bottom of the internal auditory meatus were underperformed, but the brainstem was not abnormally underperformed (14).

Figure 3. A (T1), B (T2), C (FLAIR), axial image showing long T1 and T2 signals, high FLAIR signal in the left middle peduncle cerebellaris and outside the colliculus; the left facial nerve and vestibulocochlear nerve of the cisternae swell (long red arrow). D, 3D-CISS image shows the left facial nerve and swelling and thickening of the vestibulocochlear nerve; E and F, Axial gadolinium image showing section of cisternae and internal auditory canal of the left facial nerve and vestibulocochlear nerve of cisternae.



Taken from: Long-term MRI signal change in the spinal trigeminal nucleus and tract after Ramsay Hunt syndrome and trigeminal herpes zoster.

Discussion:

Between 10 and 20% of the general population will present a picture of herpes zoster throughout life, the risk progressively increasing with age, due to a decrease in the cellular immune response against varicella zoster, so that at the age of 85, 50% of the subjects will have had herpes zoster. Multiple circumstances have been described (such as stress, illness, emotion, the administration of immunosuppressive drugs, or radiotherapy) that, by compromising the patient's immunity, can

trigger the reactivation of the virus and its proliferation through the lymph nodes. and affect several nerve branches. In comparison, facial nerve involvement (VIII) was present in 18 patients (90%), isolated or as part of a polyneuropathy. In a study by Coulson et al. Regarding the temporal sequence of clinical findings in SRH, neuralgic pain in Hunt's area was the first symptom in half of the patients, followed by facial paralysis that appeared as the first symptom in 20% and only in 2 % the first manifestation was herpetic vesicles of the external ear (9). And in the magnetic resonance we will observe an increase in the signal intensity of the canalicular segment and the geniculate ganglion to the inflammation of the nerve caused by the VVz and also in T2 sequences it is registering a thickening of the distal portion of the intrameatal nerve. In general, in SRH, there is a 50% enhancement of the facial nerve on contrast-enhanced MRI, and after contrast administration

there is also enhancement of the eighth nerve in the distal internal auditory canal and in the labyrinthine segment (10).

The relationship between a hyperintense signal seen in contrast-enhanced T1-weighted facial nerve swelling has been poorly studied to date, with the exception of a study by Kim et al. who examined 13 patients who underwent facial nerve decompression surgery through a middle cranial fossa. Approach with or without a transmastoid approach. Kim et al. It demonstrated a good correlation of contrast enhancement with facial nerve swelling in the labyrinthine and geniculate segment. (9) Magnetic resonance imaging is nonspecific as a diagnostic technique if we do not have one prior to the onset of paralysis or it is incomplete, although it is effective in assessing the prognosis of peripheral paralysis by evaluating the uptake of gadolinium contrast by the nerve, in the acoustic pore. This syndrome is the second most common cause of atraumatic peripheral facial paralysis (12).

Conclusion:

The typical characteristic findings in a Magnetic Resonance Imaging (MRI) in patients with Ramsay Hunt syndrome we will observe an increase in the signal intensity of the canalicular segment and the geniculate ganglion to the inflammation of the nerve caused by the VVz and also in T2 sequences it is registering a thickening of the distal portion of the intrameatal nerve. In general, in SRH there is a 50% enhancement of the facial nerve on contrast-enhanced MRI, and after contrast administration there is also enhancement of the eighth nerve in the distal internal auditory canal and in the labyrinthine segment. By compromising the patient's immunity, they can trigger the reactivation of the virus and its proliferation through the ganglia and affect various nerve branches. However, nuclear magnetic resonance is nonspecific as a diagnostic technique if we do not have one prior to the onset of paralysis or it is incomplete, although it remains effective in assessing the prognosis of peripheral paralysis by evaluating gadolinium contrast uptake by the nerve, in the acoustic pore.

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How to cite this article: Muñoz, A. F. D. ., Ibrahim, T. M., Ortiz, C. T. N. ., Chávez, Ángel F. L. ., Amaya, G. P. B. ., Galvis, M. C. C. ., Pacheco, M. E. F. ., & Herrera, M. A. I. . (2022). Typical Mri Findings of Ramsay Hunt Syndrome. Journal of Medical Research and Health Sciences, 5(4), 1899–1905. https://doi.org/10.52845/JMRHS/2022-5-4-7