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Case Report

Atypical Herpes Zoster Oticus: Case Report and Review of Literature

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A B S T R A C T

Introduction and Importance: Herpes zoster oticus, or Ramsay Hunt syndrome, is a rare neurological pathology caused by the varicella-zoster virus reactivating in the facial nerve's geniculate ganglion. Its importance lies in the description of a rare case of cranial nerve palsies.

Case presentation: A 19-year-old previously healthy man was admitted to our emergency department complaining of an intense right otalgia in the last two days, without improvement with simple analgesia. Symptoms were complicated by right facial palsy, mild proper hearing loss, dysgeusia, and vertigo 24 hours later. On physical examination, he was alert and oriented, with a Glasgow score of 15/15, without focal neurologic deficit except for right-sided peripheral facial palsy: Grad V of House–Brackmann scale. On otoscopy, he had no visible vesicular eruption nor any abnormalities.

Clinical discussion: Sicard syndrome, also known as Herpes zoster oticus or Ramsay Hunt syndrome, is a rare neurological condition caused by the reactivation of latent VZV in the geniculate ganglion of the 7th cranial nerve.

Clinical diagnosis is mainly based on interrogation findings and clinical examination. Three clinical pictures are possible:

1. The diagnosis is usually easy when the vesicular rash occurs before or simultaneously with the facial palsy
2. Yet, sometimes, the rash develops only after the onset of facial palsy
3. And finally, a minority of patients, like our case, do not develop any vesicular lesions at all.

Conclusion: Early diagnosis and adequate treatment by virostatic agents and corticosteroids are crucial to improve damaged nerves and maximize and hasten the chances of full recovery.

Keywords: Herpes zoster oticus; Sicard syndrome; Lagophthalmo; Ipsilateral palsy

Introduction

Ramsay Hunt syndrome or herpes zoster oticus due to varicella-zoster virus infection is commonly reported in immunocompromised individuals or patients aged at least 50. It is responsible for 16% of unilateral facial paralysis in children and 18% in adults. Its classic triad is composed of vesicle-cracked lesions in the concha, ipsilateral palsy, and lagophthalmos.

In fact, immunosuppression, advanced age, pregnancy, lack of vaccination against the varicella-zoster virus, and physical and psychological stress are major factors that predispose to the

reactivation of the virus, which may invade the central nervous system. Diagnosis is based on a bundle of arguments: clinical history and physical examination. Further investigations, such as an MRI, can be carried out to rule out differential diagnoses.

We report a case of Ramsay Hunt syndrome in a young man without any underlying pathology. This highlights the importance of early diagnosis to minimize neurological damage and emphasize that even if the clinical triad appears late or is incomplete, treatment must be early for a significant improvement in symptoms and quality of life.

This work has been reported as being in line with the SCARE criteria.

Case Presentation

A 19-year-old previously healthy man was admitted to our emergency department complaining of an intense right otalgia in the last two days, without improvement with simple analgesia. Symptoms were complicated by right facial palsy, mild proper hearing loss, dysgeusia and vertigo 24 hours later. He denied fever, tinnitus, hyperacusis, nausea or vomiting. On physical examination, he was alert and oriented, with a Glasgow score of 15/15, without focal neurologic deficit except for right-sided peripheral facial palsy: Grad V of House-Brackmann scale. On otoscopy, he had no visible vesicular eruption nor any abnormalities **(Figures 1 and 2)**.

Figure 1: Right-sided facial palsy grade 5 according to House-Brackmann facial grading scale.

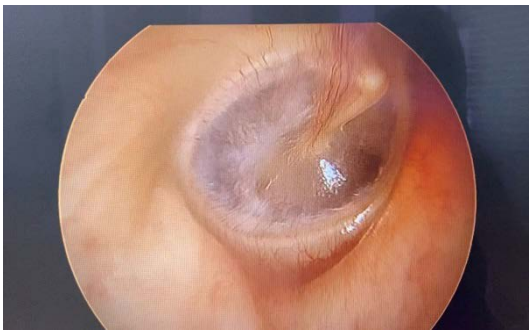

Figure 2: Normal tympanic membrane.

He also had gait ataxia and was complaining of rotatory vertigo, all of which made vestibular examination difficult. However, he had no dysmetria on finger-to-nose and heel-to-knee tests.

With the clinical findings, the diagnostic hypothesis of Herpes zoster oticus was suggested. During hospitalization, further investigations were performed. Brain MRI showed an asymmetric enhancement in the right facial nerve after contrast administration, suggesting right facial neuritis **(Figure 3)**.

Pure tone audiometry examination revealed increased bone and air conduction thresholds in frequency ranges approximating 1000 Hz and exceeding 2000 Hz on the right side, suggesting sensorineural hearing loss.

Vestibular tests showed decreased VOR gains of the right anterior and lateral semicircular canals **(Figure 4)**. Videonystagmography revealed a homogeneous pursuit and a left preponderance. In contrast, in the caloric testing, a right areflexia with a deficit of 93% was obtained **(Figures 5,6,7)**.

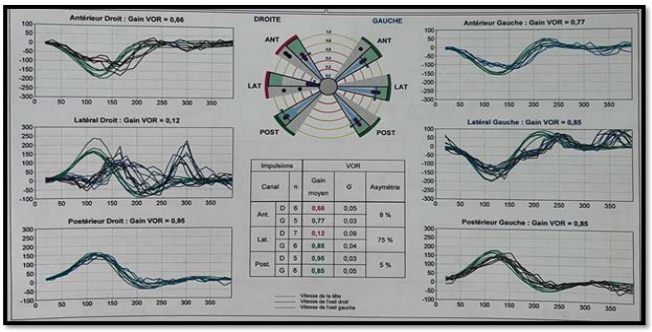

Figure 3: A video head impulse test of the patient before the treatment shows decreased VOR gains of the right anterior and lateral SCC.

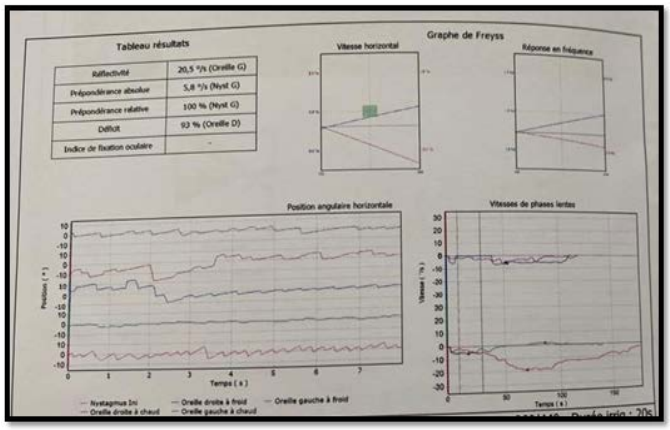

Figure 4**:** Caloric testing reveals right areflexia in low frequencies that have not yet been compensated, with a significant left preponderance.

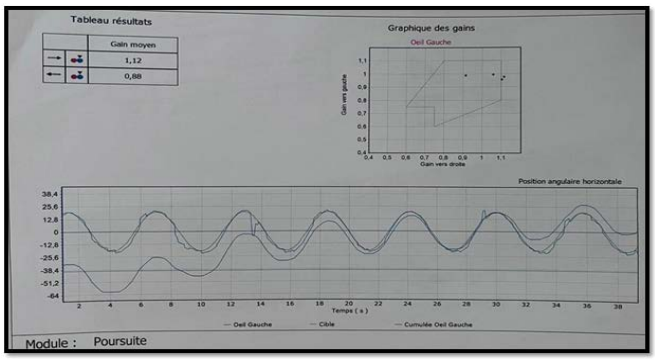

Figure 5: Smooth pursuit testing with no abnormalities.

Medication included intravenous acyclovir 10 mg per kg every 8 hours for 14 days and prednisolone 1mg/kg for five days.

Physiotherapy for the face and facial nerve rehabilitation was initiated, as well as vitamin B therapy, proper eye protection, and humidifying eye drops. Vestibular rehabilitation was also initiated during the hospitalization.

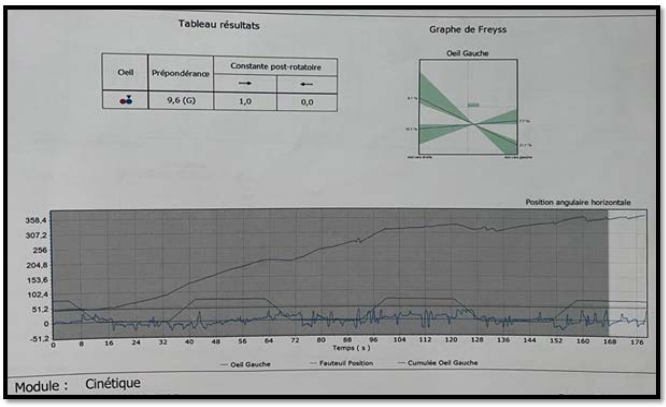

Figure 6: This test shows an important left preponderance.

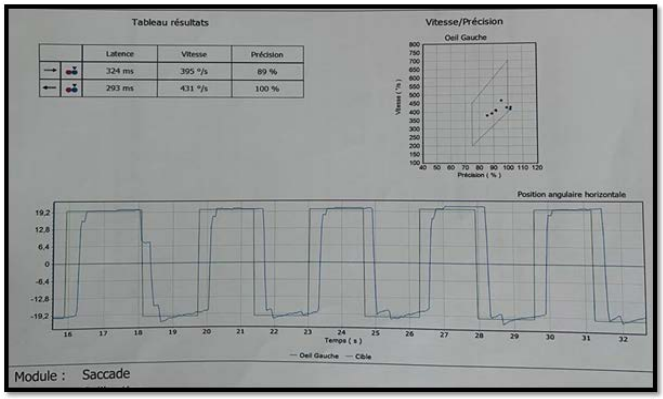

Figure 7: No saccadic disorder detected in Videonystagmography tests.

The patient slowly improved and showed complete improvement in gait. After four days, he was discharged from the hospital to complete home treatment and return to follow-ups. He had a grade III of the House Brackmann Facial Nerve scale system at discharge **(Figure 8)**.

Figure 8: Improvement of facial function after four days of intravenous treatment.

After one month, he presented partial improvement in the facial palsy. He is in grade II of the House Brackmann Facial Nerve scale system.

No vertigo, no gait ataxia was found. Control vestibular testing showed a VOR gain of the right lateral SCC of 0.30 vs. 0.12 and a VOR gain of the right anterior of 0.93 vs. 0.66.

At caloric testing, we noted a well-compensated right hypo valence and a reflective lateral SCC on both the right and left **(Figures 9,10)**.

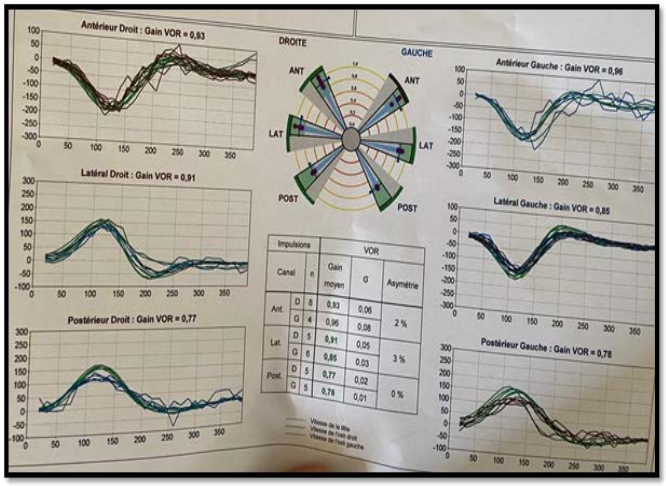

Figure 9: Video head impulse test after one month, showing normal VOR gains.

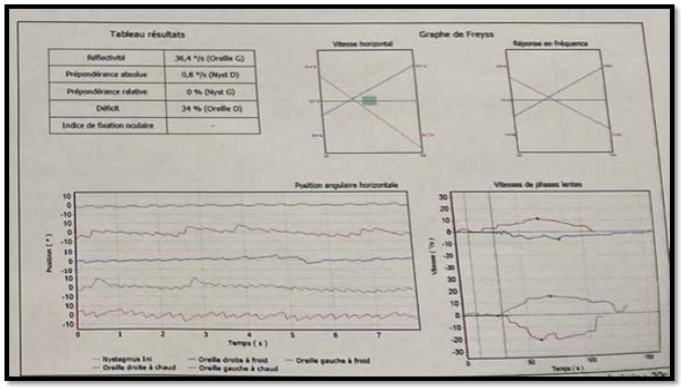

Figure 10: Control caloric testing after one month.

Discussion

Facial nerve paralysis is idiopathic in more than 70% of cases (Bell's palsy). However, other varied pathologies can be responsible for acute facial nerve paralysis: autoimmune diseases, sarcoidosis, neurological diseases, temporal bone trauma, iatrogenesis, neoplasms (glomus tumor, vestibular schwannoma, parotid), as well as Varicella-zoster virus, and cytomegalovirus infections[1].

icard syndrome, also known as Herpes zoster oticus or Ramsay Hunt syndrome, is a rare neurological condition due to the reactivation of latent VZV in the geniculate ganglion of the 7th cranial nerve[2].

The RHS was first described in 1906, and many case reports and clinical and pathologic studies have been published. However, the more is published, the less this syndrome is understood. Either because of its rarity or because of the unpredictable clinical condition[3,4].

Hunt established a classification of his syndrome in four different clinical groups[5]:

1. Herpes oticus without neurologic signs.
2. Herpes oticus with facial palsy.
3. Herpes oticus with facial palsy and auditory symptoms.
4. Herpes oticus with facial palsy with accompanying auditory and labyrinthine symptoms.

This classification was based on the hypothesis that the pathologic process of this disease is taking place at the geniculate ganglion. Yet, since the publication of this classification, many authors have questioned the validity of the geniculate ganglion theory between 1907 and 1967. Consequently, the consistent finding in autopsies was a widespread lymphocytic infiltration along the entire facial nerve, not just about the geniculate ganglion[6,7].

The incidence of zoster oticus appears less in children who receive the varicella vaccine than in children who contract varicella infection, suggesting that vaccination might prevent herpes zoster infection and prevent or reduce the occurrence of Ramsay Hunt syndrome. However, the reactivation of latent varicella zona virus is triggered by a decrease in patient cell-mediated immunity[8].

Clinical diagnosis is mainly based on interrogation findings and clinical examination. In fact, regarding the timing of the vesicular rash and the facial palsy, three clinical pictures are possible[9]:

1. The diagnosis is usually easy when the vesicular rash occurs before or simultaneously with the facial palsy,
2. Yet, sometimes, the rash develops only after the onset of facial palsy,
3. And finally, a minority of patients, like our case, do not develop any vesicular lesions at all.

Although the triad blister pustule crusted lesions in the external ear around the Ramsay Hunt region, lagophthalmos, and finally, peripheral ipsilateral facial paralysis are sufficient to make the RHS diagnosis[10]; However, clinical diagnosis can sometimes be difficult when faced with a polymorphic and dissociated presentation[11].

Other symptoms may occur, such as hearing loss, tinnitus, dizziness, and, less frequently, dysgeusia, nasal obstruction, dry eye, and dysarthria[12].

In the case described here, attention is drawn to the lack of dermatological lesions. Further tests may be performed to rule out other possible differential diagnoses. Virological and serological diagnoses are particularly interesting in the face of severe and atypical forms. Polymerase chain reaction assays may be useful to detect herpes zoster virus DNA in exudates from ear lesions or cerebrospinal fluid[13]. In addition, brain MRI can visualize inflammation of the cranial nerves[14].

Regarding treating herpes zoster oticus, intravenous administration of virostatic agents such as acyclovir in combination with corticosteroids is highly recommended. It should be administered early to improve acute pain and reduce the duration of active disease, reducing neural damage and preventing postherpetic neuralgia.

The virostatic agent acyclovir is recommended at a dose of 10 mg/kg in adults and 500 mg/m2 in children every 8 hours for a minimum duration of 7 to 10 days, followed by oral treatment of 7 days. Corticosteroid therapy is controversial; the dose is generally 1 mg/kg/day in 10 days, has potent anti-inflammatory action, and is well tolerated[15].

However, it seems that there is no difference between the use of oral versus intravenous antiviral agents[16]. What matters is starting treatment in the first 48 hours of clinical symptoms, as we did in this case, which is a crucial prognosis factor in preventing nerve damage[17].

Treatment also involves adequate analgesia and motor physiotherapy to treat motor sequelae, which tend to be more severe and less likely to improve when compared to Bell's palsy[18].

Conclusion

Ramsay Hunt syndrome is defined by zoster oticus, which involves the seventh and the eighth cranial nerves. It is characterized by acute facial palsy and vestibulocochlear dysfunction with a herpetic eruption on the auricula and external ear. Polymorphous and dissociated clinical presentations are possible, which shows the diagnosis challenges of this syndrome, especially in the absence of a typical vesicular rash.

Early diagnosis and adequate treatment by virostatic agents and corticosteroids; is crucial to improve damaged nerves and maximize and hasten the chances of full recovery. This allows us to conclude that the prognosis depends on the timing at which the combined therapy is started.

The patient provided written informed consent for the publication of this case report and accompanying images. The editor-in-chief of this journal can review a copy of the written consent upon request.

Ethics statement: Ethics approval is not needed in case reports in our institution's research structure.

Conflict of Interest: All authors have no potential conflicts of interest to disclose.

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