Varicella-Zoster infection with isolated cochleovestibular affectation (without facial palsy)

Servicio de Otorrinolaringología. Hospital Universitario de Getafe. Madrid.

Abstract: The otological complications of Ramsay Hunt Syndrome include facial palsy, tinnitus, hearing loss, vertigo, dysgeusia and rashes. The lower cranial nerves are sometimes affected by this neuritis. We present the case of a man with an unimpaired immune system that had cranial mononeuritis with unilateral involvement of the facial and vestibulocochlear nerves after infection with varicella-zoster virus, without herpetic lesions.

Key words: Ramsay Hunt. Varicella-zoster virus.

INTRODUCTION

The varicella-zoster virus can cause otological complications such as facial paralysis, hearing loss, vertigo, dysgeusia and a rash.

We present a case with Ramsay Hunt syndrome that had the classic characteristics, (sensorineural hearing loss and vertigo from affectation of the vestibulocochlear nerve), except that there was no peripheral facial paralysis; only the sensitive part of the facial nerve was affected, which is typical in pediatric and young patients, but not common in adults. The diagnosis was confirmed with serology tests.

The aim of this article is to raise awareness in order to facilitate the early diagnosis of other cases.

Anamnesis

A 59-year old male patient who had rotatory vertigo for two days accompanied by a neurovegetative reaction associated with a feeling of aural fullness, sudden hearing loss in the left ear and otalgia. The patient did not complain of otorrhea or tinnitus. The patient had no antecedents of infectious symptoms of the upper airways. At first he did not present symptoms of paresthesia or facial paralysis, but 10 days later he had a tingling sensation in the left half of his face without motor affectation. There was no other neurological focal sign throughout the evolution of the clinical picture.

The patient had been attended to in the emergency department the week before for otalgia and tumefaction of the left pinna, hearing loss and nausea, and was diagnosed with perichondritis and acute otitis media; medical treatment was prescribed.

Of his personal antecedents, only the fact of his being an ex-smoker stands out. He did not have high blood pressure, diabetes or any other systemic illness. There was nothing in the family medical history of any significance.

Physical exploration

Otoscopy: Intact and normal tympanic membranes with vesicular scabs on the left pinna (concha) and the entrance to the external auditory canal.

In the neurological exploration the patient presented a grade II Alexander spontaneous horizontal rotatory nystagmus to the right that intensified with Frenzel's glasses. The Nylen-Bárâny directional test: preponderance to the left. Unterberger test: positive, turning to the left. Romberg's test with eyes open: stable, with eyes closed: unstable. Cranial nerves, including the facial nerve: normal. The rest of the ENT examination was without anomalies.

Complementary tests

Pure-tone audiometry (Figure 1) showed normal hearing in the right ear and moderate sensorineural hearing loss with PTA of 48dB in the left.

Given the asymmetry of the audiometry, BAEP was conducted: with symmetrical, non-pathological values for waves I, III and V and with a non-pathological inter-peak difference of I-V in relation to the control group of the same age.

In the visual-ocular tests, the following results were obtained: saccadic follow-up with low gain, normal saccades and symmetrical optokinetic. The caloric and
bithermal tests were done when the otic lesions the patient presented improved; normal labyrinthine excitability and the absence of directional preponderance were obtained at the time of the recording.

The computerized dynamic posturography (CDP) showed a severe pattern (multisensorial). The vestibular autorotation test (VAT) showed left asymmetry of the vestibular-ocular reflex (Figure 2).

Clinical impression and treatment

With all the above data, a diagnosis of infection from varicella-zoster was reached, with affection of the left vestibulocochlear nerve and the sensitive part of the VII pair of the same side. The patient was treated with 800 mg/4 hours of acyclovir, 5 doses a day for 10 days; sulphpride: 50 mg/8 hours for 5 days; and metamizol magnesium: 1/8 hours, substituted by carbamazepine 200mg/12 hours for 3 months for post-herpetic neuralgia due to affection of the sensitive part of the facial nerve. Vestibular rehabilitation through computerized dynamic posturography and optokinetic stimulation was prescribed to improve postural control disorders when standing and the vestibular-ocular reflex.

Immediately after rehabilitation the VAT continued to show low gain at high movement frequencies and a phase advance in all frequencies. The CDP showed a slight improvement, with a visual-vestibular pattern (Figure 3).

Evolution

In the check-up conducted one month later, the patient felt unstable with a blocked up feeling and
tinnitus in the left ear accompanied by pain and a burning feeling. Another audiometry was conducted with the same results as the first.

Instability persisted 6 months later. As recovery was slow, we suspected the virus had produced some kind of encephalitis of the brainstem, which was restricting the central compensation mechanisms. Vestibular rehabilitation was necessary and we added trimetazidine at 1/8 hours for 6 months to aid vestibular compensation.

In the check-up after a year the patient felt much better, only reporting occasional nausea with head movements accompanied by sporadic tinnitus in the left ear. The following tests were conducted: a VAT that revealed slight left asymmetry at high frequencies; a normal CDP (Figure 4); and a normalized audiometry. Given the improvement, the patient was discharged.

DISCUSSION

This combination of three symptoms: facial paralysis, auricular herpes, (vesicles or scabs in the distribution area of the facial nerve), and hearing loss was described by Ramsay Hunt in 1907 as a new syndrome. Ganglionitis of the geniculate caused by herpes zoster virus (the primary infection manifests itself as chickenpox, remains inactive in the geniculate ganglion and later reactivates: zoster) was postulated as the physiopathological cause. The auditory symptoms are explained by the proximity between the inflamed geniculate ganglion and the acoustic nerve.

Later, in 1944, Denny-Brown described the affectionation of other cranial nerves and suggested that there were subgroups of the new syndrome. In 1967, Blakely suggested that the obsession with facial paralysis obscured the extension of the process, which should be known as cephalic herpes zoster.

These days the syndrome is recognized as multiple peripheral neuritis, characterized by the affectionation of both sensitive and motor nerves. The facial and acoustic nerves are the cranial pairs most often affected because of their anatomical proximity in the inner auditory canal; the trigeminal, vagus and the glossopharyngeal nerve can also be affected; this can be explained by their being close to each other, stemming from the same branchial arch (VII, VIII and IX) or transaxonal across the vaso vasorum that supply the nerves.

The neurological complications of herpes zoster are numerous; post-herpetic neuralgia being the most common among them. The next most common complication is peripheral motor neuropathy, followed by a wide variety of syndromes that include cranial paralysis, myelitis, encephalitis, thrombotic cerebral vasculopathy, acute ascending polyradiculitis and aseptic meningitis.

The incidence of Ramsay Hunt Syndrome is of 5 cases per 100,000 inhabitants and per year in the USA. Epidemiologically, segmental paresis provoked by infection from varicella zoster virus predominates in middle-aged and elderly patients; motor affectionation is rare in children and young adults, but when it occurs it is localized in the cranial muscles.

Vertigo, a well-known manifestation of the Ramsay Hunt syndrome, is a factor of bad prognosis in regard to the speed of recovery from hearing loss.

Abramovich and Prasher reported vertigo in 11 of 13 patients (85%) with Ramsay Hunt syndrome, while Devriese indicated the presence of vestibular symptoms in 23 of 32 patients (72%), Blackley et all demonstrated the histopathological affectionation of the modiolus and vestibular nerves. Proctor et al described labyrinthine destruction and vestibular dysfunction in one patient with infection from varicella zoster virus.

Byl and Adour described the different types of hearing levels found in patients with idiopathic facial paralysis or herpes zoster. They found sensorineural hearing loss with facial paralysis in 11 of 172 patients, that is to say 6.5%. Severe or profound hearing loss was found in 3 of the 11 cases. Satisfactory hearing recovery occurred in patterns with cochlear lesions with moderated sensorineural hearing loss.

Fumihisa Hiaride et al compared the hearing levels in patients with Ramsay Hunt Syndrome and hearing affectionation with others without hearing affectionation. Half the patients presented moderate sensorineural hearing loss in high-pitched sounds and recruitment in 6 of the 8 patients. They speculated that the herpes zoster virus affects the sensory cells of the organ of Corti to a greater degree than the spiral
ganglion in the cochlea or the acoustic nerve in the inner ear canal. Most of the patients recovered spontaneously from this hearing loss.

In our case, we present a patient with Ramsay Hunt syndrome with the classic characteristics, (sensorineural hearing loss and vertigo caused by affection of the vestibulocochlear nerve), but with the uncommon absence of peripheral facial paralysis, affecting only the sensitive part of the facial nerve, typical of pediatric and teenage patients, but not of older ones. The diagnosis was confirmed with blood tests, the exploration, and by the results of the complementary tests conducted.

CONCLUSION

In conclusion we can say that the Ramsay Hunt Syndrome presents broad clinical variants which have the diverse affection of certain cranial nerves in common. In such cases blood tests should be conducted to confirm the suspected diagnosis.

References