Letter to the Editor

Facial diplegia resembling bilateral Ramsay Hunt Syndrome

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VZV

1. Introduction

The simultaneous and bilateral peripheral paralysis of the facial nerve is defined as a paralysis that affects both sides of the face within 4 weeks. It is an extremely rare condition and it occurs in 0.3 to 2% of patients with unilateral facial nerve paralysis [1] and is less than 1% as frequent as the unilateral form [2], having an annual incidence of 1 case for 5,000,000 people [3].

Here, we describe a case of a young patient with a series of neurological symptoms suggestive for Ramsay-Hunt Syndrome sine herpette.

2. Case report

A 23-years-old woman, with recent temperature (38.5 °C), arrived to our emergency room with bilateral facial muscular deficits, more noticeable on the right side, from at least two days and with a worsening trend.

In the previous five days, she reported self-limited vesicular rash in hard palate and the anterior two-third of the tongue treated with anti-inflammatory therapy and in remission when she has come to our attention.

She had been properly vaccinated in accord to the Italian schedule of vaccination. There were no clinical history, signs and symptoms of immunosuppression.

At hospital admission, we performed a blood count which showed normal blood chemistry values, except VES 45 mm/h (normal value 2–30 mm/h), positive anticardiolipin antibodies (type IgG: 24.7 U/ml), IgG anti HSV-1 slightly positive, IgG anti VZV 2269 mIU/ml (normal values until 165 mIU/ml), lymphocyte population was at 17%.

Computer Tomography scan was negative.

The neurological examination documented bilateral facial paralysis, more evident on the right side than left, associated with incomplete closure of the eyelid rhymes and bilateral Bell's sign. She referred hypoesthesia of the hard and soft palate; also, reported occasional tingling charged to the face. No further neurological signs had been found.

Magnetic Resonance (MR, day 2) documented a minimum thickening and slight contrast-enhancement, that affected the end of the internal auditory canal bilaterally, most noticeably on the right side where it also involved the vestibule and the semicircular canals; moreover, the contrast-enhancement interested simultaneously and symmetrically

the intra-canalicular segment of the facial nerve and geniculate ganglion on the right side (Fig. 1).

Therefore, we suspected that patient had either viral infection or a paraneoplastic neurological syndrome so we performed a PET/TC, which resulted negative, and a lumbar puncture which showed normal values except for serum glucose at 141 mg/dl (normal value 65–100 mg/dl).

As a result, we decided to administer a therapy with acyclovir, ranitidine, dexamethasone and paracetamol.

Neurological symptoms improved during the treatment and at discharge (day 6) remained a moderate deficit in upper and lower facial muscles, tactile hypoesthesia in the right maxillary region and tongue.

3. Discussion

In 1907 J. Ramsay Hunt described a syndrome characterized by acute facial palsy, eight nerve dysfunctions in association with a herpetic eruption on the auricle and external ear canal [4]. This syndrome was attributed to infection by Varicella Zoster Virus (VZV), which becomes latent in sensory ganglia (geniculate, vestibular and spiral) after a primary infection and emerges from latency to cause Zoster in adults [5] and it was called Ramsay Hunt Syndrome. However also Herpes Simplex Virus (HSV) could be the cause of a Ramsay Hunt-like syndrome [6].

Usually, bilateral peripheral paralysis may appear as one of the symptoms of a system disease that requires urgent treatment, such as Lyme disease, Guillain-Barre syndrome, leukemia, sarcoidosis, bacterial meningitis, syphilis, Moebius syndrome, infectious mononucleosis, skull fractures [1,3] and paraneoplastic neurological syndromes.

A differential diagnosis is the Bell's palsy, which is usually unilateral and it might mimic herpes zoster sine herpette [7] and usually doesn't cause a consensual involvement of the eighth nerve. Also, the main causes of Bell's palsy are linked to forms of *frigore*, by HVS, VZV, as well as by autoimmune syndromes. It is essential in these cases the analysis of the antibody profile in particular an elevation of the title IgG and IgM anti VSV or an increase of 4 times of the total of anti VZV antibodies title [8].

The VZV, as the HSV, are virus of the family of *Herpesviridae* and are both neurotropic virus which could be latent in the geniculate, vestibular and spiral ganglia and could reactivate, during a period of immunosuppression, and cause neurological symptoms and signs.

But it could be cases of Ramsay-Hunt syndrome sine herpette; some authors have suggested that patients with facial paralysis without vesicular eruptions on the face can be diagnosed with the syndrome of Ramsay-Hunt if they showed an increased antibody titer of anti-VZV or cochlear-vestibular signs [9,10].

In our case the trigger factor, for the described case of bilateral Ramsay-Hunt syndrome, may be, most probably, a viral infection caused a reactivation of VZV also in consideration of higher titer of IgG anti VZV.

Usually facial weakness reaches its peak by 2–3 weeks; complete paralysis occurs in 67% of patients and it is more common in older patients. Immunosuppression and multiple cranial nerve involvement also

Fig. 1. MR examination pre (A, B, E) and post (C, D, F, G) contrast medium. Open arrow: enhancing of geniculate ganglion and the ipsilateral great petrosal nerve (C, D). In F is showed the enhancing of the right vestibule and semicircular canals (arrow); also, note enhancing of upper part of auditory-vestibular-facial nerves corresponding to facial nerve (close arrows on G).

adversely affect outcome. Complete recovery occurs in 67% of patients with incomplete facial paralysis and only in 10% of those with complete paralysis [11].

4. Conclusion

Our case of bilateral Ramsay-Hunt syndrome could be caused by reactivation of VZV (also in relation increase of 14-fold of IgG anti-VZV). This condition should be treated, as soon as possible, with systemic steroids and acyclovir which could prevent the incidence of

permanent facial palsy and should be given every time when relapse occurs.

Conflicts of interest

None.

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Author contributions

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