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A Case of Recurrent Facial Palsy Associated with Anti-GM2: is it anyway Guillain-barré syndrome?

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ABSTRACT

Objective: A rare case of possible hereditary predisposition to autoimmune neuropathy

Background: This case report is of a patient who presented two episodes of Facial paresis in her live without ascertained apparent causes. Bell's palsy is commonly known as peripheral idiopathic facial nerve palsy, because in the most cases the triggers remain unknown (1).

Case Report: I want to present a case of a 34 year old woman who we will call A.B. and who came to us to evaluate the severity of the sequelae of a Bell´s palsy and any eventual need for cosmetic measure. But she had another Bell´s Palsy 9 years ago, so I started to investigate the most common and treatable causes of Facial paresis. I found a significate increase of anti-bodies against ganglioside GM2. When I explained to her that these anti-bodies is commonly related with several neurological diseases, she started to investigate her family history and she found that his father's brother died of Guillain–Barré syndrome about 40 years ago.

Keywords: Bell´s palsy, facial, palsy, paresis, Guillain–Barré syndrome, ganglioside, Antibodies, GM2.

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Background

Facial Paresis is more commonly idiopathic (Bell´s palsy), there are many factors that can cause a facial paresis such as some infections, e.g. Herpes simplex, Varicella Zoster, Chlamydia pneumoniae, HIV, Epstein-Barr virus, Campylobacter, rickettisia, cytomegalo-virus, rubella virus, Borrelia. But in other cases, a facial paresis can be a part of a more complex syndrome, e.g. sarcoidosis, Sjögren syndrome, Merkersson-Rosenthal syndrome, Wegener disease. Another group of facial paresis is caused by an anatomical injury of the nerve, e.g. petrous bone fracture, tumors (especially parotid carcinoma). In which case, a facial nerve paresis can be the debut form of generalized polyneuropathies such as Guillain–Barré syndrome or Miller-Fisher syndrome. To assess the severity of facial paralysis and to estimate the prognosis, the House and Brackmann scale is commonly used. The treatment of choice in the acute phase is glucocorticoids for 10 days, more commonly Prednisolone.

Case Report

This case report concerns a 34 year old woman of North European Caucasoid race who we will identify in this report as A.B. who came to us to evaluate the severity of the sequelae of a Bell´s palsy and any eventual need for cosmetic measure. I met her for the first time exactly 116 days after the first symptom of her second episode of Bell´s palsy. She had her first Bell´s palsy exactly 9 years before, in the same month, that was June. Before both episodes, she had a subjective paresthesia on the left, around the mouth and in the nasolabial groove, in the first episode she can also remember a pain sensation behind the ear on the same side. Both episodes enveloped the left facial nerve. According to the ambulance report, in both cases all monitoring vital parameters were within the normal range (2011: SpO2 99%, heart rate 81/min, Blood pressure 129/61mmHg, temperature 36,8. 2020: respiratory rate 18/min, heart rate: 80/min, blood pressure 120/60mmHg, temperature 36,6). In the first episode there was the suspection that the vaccination against the swine influenza (H1N1) could have acted as a trigger, but in the second episode no triggers had been found. In fact, an otitis 15 months earlier (2019) and IgM-type antibodies at borderline levels against Borrelia in the serum, 4 months later, cannot be considered reasonable triggers. In both cases, she couldn´t remember any form of rash or tick bites, she never had migrant arthritis or symptoms attributable to Lyme Borreliosis. The absence of a rash and neuropathic pain makes VZV and HSV acute infections or exacerbations implausible. She denies head trauma which could cause a fracture of the temporal lobe. A.B. remembers that she had mononucleosis and varicella in childhood but doesn´t remember herpetic infections. Except hair loss, the history and physical examination totally exclude Parry-Romberg syndrome. The absence of bulbar symptoms such as the loss of the swallowing reflex excludes Kennedy syndrome. A.B. denies nausea, deafness, tinnitus and diplopia, but she remembers vertigo, this symptom can make one suspects the Ramsay Hunt syndrome. On the other hand, the first doctor who examined her at the emergency room was a otolaryngologist specialist who found healthy ears on otoscopic examination, and who valued this as a possible Bell´s palsy in progress. A.B. had an idiopathic fever for 3 days, fluctuating between 37,9 and 38,3 exactly 5 weeks before the debut of the facial palsy. The origin of the fever is unknown and without any other symptoms, but this was reasonably viral. This anamnestic finding removes the doubts regarding the genuineness of the diagnosis of Bell´s palsy.

9 years earlier the Serum protein electrophoresis had shown an increase in ceruloplasmin, which can be interpreted as an activation of lymphocytes. A.B. was not treated with corticosteroids, the Sunnybrook Facial Grading System showed a worsening from 41 points composite score 11 days after the debut (5 resting symmetry score + 44 Voluntary movement score + 0 Synkinesis score) to 81

points composite score 34 days after the debut (5 resting symmetry score + 76 Voluntary movement score + 0 Synkinesis score). The search for IgM and IgG antibodies against Borrelia in the CSF and serum was carried out the same day of the debut of the Bell´s palsy and was negative.

After the first Bell´s palsy, A.B. recovered all the function in about 6 months despite the absence of corticosteroid treatment. This means that any functional deficit at the debut of the second Bell´s palsy was new.

At around 8 pm on the day of the debut of the second episode with Bell's palsy, it was evident that the smile was not symmetrical, the left eyebrow and eyelid were slightly lowered. 4 days later A.B. started treatment with 60mg Prednisone/day and 5 days with dose reduction of 10 mg each day. 116 days later after the first symptom, A.B. didn't have Bell phenomenon and crocodile tears sign, nor Jaw-winking phenomenon. She didn't have synkinesis. The tropism of the face was normal and symmetrical. No swelling or redness plication of the tongue. The House and Brackmann scale showed a mild dysfunction grading II, with normal finding at rest reduced forehead innervation, nearly normal lid closure and nearly normal mouth innervation. A.B had ageusia for the sweetness of the anterior two thirds of the tongue on the left. Apparently, not only the facial nerve presents aberrations, the protopathic and epicritic sensitivity of the three branches of the left trigeminal nerve is reduced according to the patient's subjective responses. Also, the left soft palate lifts less than the right, in my opinion. The tendon reflexes were normal. In view of these atypia, and the fact that only around 8 percent of Bell's palsy cases recur over the course of a lifetime (2), I investigated further.

Chest X-ray ruled out sarcoidosis. Erythrocyte Sedimentation Rate was normal, and C-Reactive Protein levels were also normal, this excludes inflammatory processes. Glycated hemoglobin was normal, this excludes Diabetes as a cause. Antibodies against Extractable Nuclear Antigen (Inclusive SS-A and SS-B) were negative, this excludes Sjögren syndrome. Proteinase 3-Antineutrophil Cytoplasmic Antibodies were negative, this excludes Wegener´s granulomatosis. Antibodies against Aquaporin 4 were also negative. Epstein-Barr virus IgG type antibodies positive, Herps simplex type 1 and 2 antibodies type IgG positive and Antibodies type IgG positive and type IgM negative against Varicella Zoster virus, like as with memory immunity for Varicella, mononucleosis, and herpes. Antibodies against the human immunodeficiency viruses (HIV) were also negative. After nearly 17 weeks, antibodies against Borrelia were negative type IgG and borderline levels type IgM, after consultation with the infectious disease specialist, this finding was considered a cross reaction with another antigen. A.B. denies Lyme disease symptoms and tick bites. Finally, all antibodies (IgM and IgG) against gangliosides were negative (<20%), except antibodies IgM against ganglioside GM2 (47%). It is possible to imagine that this percentage would have been much higher if the examination had been conducted on the cerebrospinal fluid, at the time of the palsy and in the absence of corticosteroid therapy. When I told this to A.B. She did an investigation of her family history; it results that her father´s brother had died of a fulminant Guillain-Barres syndrome about 40 years ago.

Discussion

A facial palsy is more often idiopathic [1] and non-recurrent [2]. Causes of recurrent facial palsy are Melkersson-Rosenthal syndrome [1], dehiscent facial nerve canal, tumors [5]. Moreover, recurrence in patients with idiopathic facial palsy may increase the likelihood of recurrence and worsen prognosis [5].

Recurrent Facial palsy have just a few know comorbidities risk factors, such as diabetes and hypertension [4]. On the other hand, there are a few known factors which can explain higher risk to get sick with Guillain-Barre syndrome, such as immune-mediated and rheumatological disease, cancer, transfusion, surgical procedures, and

preeclampsia [3]. Greco et al. (2012) had postulated the hypothesis that Bell´s palsy can be a mononeuritic variant of Guillain–Barré syndrome, a neurologic disorder with recognized cell-mediated immunity against peripheral nerve myelin antigens [6]. In fact, there are few cases in adults and children that debut with unilateral facial paralysis, which are ultimately ascertained as Guillain-Barre syndrome [7-8]. To reinforce this hypothesis, there is a temporal incidence correlation between Bell's palsy and Guillain–Barré syndrome at various times of the year [9].

Immunological aberrations such as elevation of several cytokines such as IL-1, IL-6 and TNF-α has been demonstrated during idiopathic palsy of the facial nerve [10]. At the same time, it is demonstrated an inflammation response and a switch from a T-cell-mediated immunity to a humoral-mediated immunity with B-cells prevalence during idiopathic palsy of the facial nerve [11,12]. If such antigens/infections can orientate to develop a Bell´s palsy instead of a Guillain–Barré syndrome is not known, but for example my patient received a vaccination against pandemic influenza A [H1N1] so called Swine influenza in 2011 and after this she developed her first Bell´s palsy. This vaccine was associated with higher risk to develop Bell´s palsy but not higher risk to develop Guillain–Barré syndrome [13]. During a Guillain–Barré syndrome several antibodies against gangliosides were found and these can orientate different clinical presentations, inclusive of facial weakness [14, 15, 16].

There are a few known cases with acute monophasic peripheral neuropathy or cranial neuropathy with isolated IgM-type anti-GM2-antibody positivity comprised a heterogeneous group of syndromes, including two cases of acute motor axonal neuropathy, one of acute inflammatory demyelinating polyneuropathy, and one of isolated facial diplegia with right side debut and Distal paresthesia. This patient was also treated with corticosteroids, and the infectious disease investigations inclusive for Cytomegalovirus, Epstein-Barr virus, Herpes simplex virus, Mycoplasma pneumoniae were negative despite the fever and upper respiratory symptoms [17]. A pediatric case who developed a classic ascending flaccid paralysis with involved cranial nerves and with isolated positive anti-GM2 IgM antibodies was also reported [18]. A cross-reactivity between GM2 gangliosides and cells which exposure Cytomegalovirus antigens is proved in vitro [19]. But not all patients with Cytomegalovirus infections and positive Anti-GM2 develop a Guillain–Barré syndrome [18], this data is in favor of a hypothesis of genetic predisposition. Mycoplasma pneumoniae infections seem also related with Anti-GM2 antibodies and Guillain–Barré syndrome [17]. A genetic predisposition to develop facial paralysis is reinforced by the existence of families with several affected members [20, 21]. The involvement of HLA genes is unclear in determining recurrent episodes of Bell´s palsy [20, 21]. But my research of the literature has found just a result of missense gene mutations of Chondroitin beta-1,4-N-acetylgalactosaminyl-transferase-1, which is in common between polyneuropathies and Bell´s palsy [22].

Conclusion

The hypothesis that Guillain–Barré syndrome and immune-mediated facial palsy are related or even two variants of the same disease is reinforced of this case report and of the current literature. One values that Anti-GM2 IgM antibodies are related not only with Guillain–Barré syndrome but even with immune-mediated recurrent facial palsy, and this report is a favorable evidence.

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