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Facial variant of Guillain-Barré Syndrome in a patient, days after being vaccinated for influenza A

Variante facial del síndrome de Guillain-Barré en un paciente, días después de vacunarse de la gripe A

Dear Editor:

Simultaneous bilateral facial paralysis is an uncommon neurological manifestation; a possible case is Guillain-Barré syndrome (GBS).. Although the facial nerve is often involved in this syndrome, it is rare for its only manifestation to be bilateral facial palsy. There are regional variants of GBS, such as Miller-Fisher syndrome, lumbar plexopathy, pharyngeal-cervical-brachial weakness, and facial diplegia.[2]

In 1976, the national influenza immunization programme in the United States was suspended after an increase in the number of cases of GBS was reported. A subsequent epidemiological study revealed a 4.0 and 7.6 relative risk at 6 and 8 weeks post-vaccination, respectively, with a risk of less than one per 100,000 vaccinations.[3] Some later works failed to reveal this increased risk.[4] Insofar as the data on the H1N1 influenza vaccine are concerned, the adverse effects reported in USA as of December 30th, 2009, were as follows: 37 GBS (with 99 million doses distributed, although the number of doses actually administered is not known).[5]

We report a case of a patient who suffered the facial variant of GBS days after having been vaccinated against influenza A.

A sixty-year-old male with a history of high blood pressure, renal colic, acute myocardial infarction in 2005 and placement of a cardiac stent in 2007. He consulted in December for moderate lumbar pain. The pain was increasing in intensity and radiated toward his waist. Five days later, he presented difficulty speaking, eating and moving his lips. These symptoms grew worse over the course of three days. The neurological examination revealed moderate bilateral peripheral facial palsy. Examination showed the remaining cranial nerves to be normal. Muscle balance and sensitivity were normal in the upper and lower limbs; osteotendinous reflexes were abolished. His gait was normal.

The following testing procedures were performed: CT and cranial NMR, chest X-ray, NMR of the lumbar spine and ECG, all of which were normal. The most striking results of the general work-up: GGT 105 U/l; 14,400 leukocytes; with normal VSG and PCR. The serological tests for mycoplasm, HIV, syphilis, Epstein Barr, CMV, *Borrelia* and *Brucella* were negative. A spinal tap was performed in which the cyto-biochemical analysis detected elevated proteins (137 mg/dL), with 4 cells; the bacteriological culture and PCR (herpes simplex, herpes zoster, enterovirus, CMV) and CSF were negative. A neurophysiological study was also performed which revealed very little alteration (responses in the blinking reflex, with stimulation of both supraorbital nerves, low amplitude desynchronization, with normal direct responses in both facial nerves).

When the patient was re-interviewed, he commented that he had been vaccinated for influenza A two weeks prior to the onset of symptoms. After receiving the vaccine, he presented intense, flu-like symptoms for 3 days (his lumbalgia and facial weakness began 14 and 18 days after having been vaccinated, respectively). He began to improve one week after maximum deficit, without treatment. He was asymptomatic one month later and the neurological examination was strictly normal.

Facial diplegia accounts for 0.3-2% of all facial paralyses and is defined as paralysis affecting both sides of the face in a period of time of no more than 4 weeks. The most common aetiologies of this bilateral involvement are: bilateral idiopathic Bell's palsy (23%), GBS (10%), multiple cranial neuropathies, multiple sclerosis, Parkinson's disease, meningeal and brainstem tumours (21%), infections (*Borrelia*, syphilis, HIV, herpes virus, mononucleosis, etc.), congenital causes (Moebius syndrome, myopathies),

porphyria, sarcoidosis, amyloidosis, leukaemias, lymphomas, diabetes, ethylene glycol intake and alcoholism.[1,6]

GBS is an acute, autoimmune, inflammatory, demyelinating polyneuropathy that typically presents as progressive, symmetrical, ascending muscle weakness. However, it may present in other ways, such as ataxia, bilateral weakness in the hands and arms, or the cranial nerves may be affected at onset, with a descending pattern of paralysis. Facial nerve involvement is common and bilateral facial palsy is characteristic. Nevertheless, peripheral facial diplegia is a very rare form of presentation (< 1%)[1] and, in general, the symptoms that are secondary to CMV infection are the ones that present in this course of illness (35% of the cases present anti-CMV IgM antibodies), with upper airway infection syndromes.[7,8] The patient we report here presented a negative blood test for CMV, although he had suffered flu-like symptoms accompanied by respiratory symptoms following vaccination.

In the series of patients with the facial variant of GBS published by Susuki[7], the most common initial symptoms were paraesthesia affecting the limbs, with facial palsy appearing 3-10 days later. In the case reported here, the initial symptom was lumbar pain, with bilateral facial weakness presenting 5 days later; we have not seen lumbalgia reported in the literature as the form of debut of this facial variant of GBS.

The association of bilateral facial weakness and hyporreflexia should be grounds to suspect the presence of GBS.[7,2] Cases of bilateral facial palsy associated with hyperreflexia due to this syndrome have also been published; in these cases there were anti-*Campylobacter jejuni* antibodies.[9] In such cases, the existence of hyperexcitability of the motor neurons and dysfunction of the inhibitory spinal interneurons has been postulated. Albumin-cytological dissociation also supports the diagnosis of a variant of GBS. Upon examination, our patient exhibited arreflexia and there was albumin-cytological dissociation in the CSF; both findings clearly uphold the diagnosis of GBS.

The reason why some cases present general or focal involvement is still moot; it has been said that there may be antigenic differences in the peripheral nerves or in their endothelium;[10] it may perhaps simply reflect the severity and duration of the underlying immunological process. Most of the cases published in the literature have had a good prognosis,[7] which is highly valuable information when contemplating possible treatment for these patients. Our patient displayed a highly favourable course of illness and no treatment measure was needed. Generally speaking, in the cases reported, the usual treatment consisted of intravenous immunoglobulins, with good response.[2,7,8]

On the other hand, influenza vaccination is associated with a significant, albeit small, increase in the risk of developing GBS; the relative incidence of GBS in the risk interval (from 2 to 7 weeks following immunization) versus the control interval of time was 1.45 in a study that was carried out in Ontario following an immunization programme.[11] In any case, it is difficult to establish the risk of GBS associated to vaccination. Although the study that was conducted following the influenza immunization programme in the USA in 1976 detected a risk of 4.9 to 11.7 per million adults vaccinated (within the first 6 weeks of vaccination), the registries that have been made subsequently have not demonstrated any relation between vaccination and GBS.[4,12] There is no answer as yet as to the cause of this increase in cases after vaccination in 1976 (and to lesser degree, after the vaccinations in 1992-1993 and 1993-1994). Some investigations suggest that this influenza vaccine might contain "contaminating" structures (such as *Campylobacter jejuni* antigens that mimic human gangliosides or other components of the vaccine) that provoke an anti-GM1 response in susceptible patients.[13] A work published in 2009 by the *Centres for Disease Control and Prevention* (United States) reviewed all the adverse events recorded for the flu vaccine between 1990 and 2005, the most common one being GBS (0.70 per million vaccinations).[14] In the case we report, the neurological symptoms appeared during the "risk" period (between weeks 2 and 3); he had not suffered any infectious process prior to or following the vaccination that could be related with GBS.

In conclusion, bilateral facial palsy requires the consideration of GBS among the differential diagnoses. The abolition of the osteotendinous reflexes and the albumin-cytological dissociation in the CSF helps to establish the diagnosis. The prognosis is, for the most part, good (even without treatment). Factors in the patient's history such as influenza vaccination must be taken into account as possible aetiology for GBS.

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Macropsia, micropsia, allesthesia, and dyschromatopsia after occipital intraparenchymal haemorrhage

Macropsia, micropsia, alestesia y discromatopsia tras hemorragia intraparenquimatosa occipital

Dear Editor,

Optical illusions are alterations in visual perception that can be characterized by an apparent modification of the size and shape of objects (dysmetropsia or metamorphopsia), seeing multiple images in the presence of a single object (polyopsia), preservation of visual images once the object that has caused the images has disappeared (palinopsia), and transposition of an object from one visual field to another (allesthesia). They may present temporarily during epileptic seizures, migraine, encephalitis, poisoning, and in psychiatric illnesses.[1] In most cases, focal brain lesions cause visual field defects. Optical illusions resulting from focal brain lesions are unusual[2]. In the review carried out, we have not found any previous case reports of combined visual illusions without campimetric alterations due to a vascular brain lesion.

A sixty-four year old female presented at the Emergency Department complaining of visual disturbances over the course of the previous 36 hours, of sudden onset, preceded by intense headache. The patient reported a mono and binocular visual syndrome consisting of three types of alterations. Firstly, and most strikingly, she reported constant variations in the shape of objects and people or metamorphopsia, such that they appeared to be extremely long and thin (macropsia), short (micropsia), wide, etc. Secondly, she reported altered colour perception (dyschromatopsia), such that objects changed in colour or even in intensity, and, finally, she described transposition of objects from one side to the other (allesthesia). The rest of the neurological examination failed to reveal any kind of focality or campimetric disturbance. A cerebral CT was performed (fig. 1) and revealed the existence of a right occipital intraparenchymal haemorrhage. Underlying lesions were ruled out by cerebral MR (fig. 2) and angio-MR. In the light of the possibility of occipital lobe epileptic seizures and despite the fact that the waking electroencephalogram failed to reveal any alterations, a decision was made to initiate treatment with levetiracetam 1000 mg/day. After 24 hours of treatment, the visual symptoms remitted and the patient was asymptomatic.

Cortical visual disorders may arise as a consequence of injury to the calcarin cortex that alters primary visual function or of lesions in associative visual areas. Permanent optical radiation and primary visual defects affect the visual field. Patients with lesions in this area may report visual

Figure 1 Cerebral CT revealing the presence of an on-going right occipital haematoma.