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Psychogenic convergence spasm mimicking ocular myasthenia

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Abstract. – A 14-year-old girl presented with a two-years history of fluctuating convergent strabismus, diplopia, and reading difficulty. She has been previously diagnosed by experienced neurologists as having ocular myasthenia and she had been treated for two years with anticholinesterase inhibitors and immunomodulatory drugs. After a thorough medical interview and neurological examination, a diagnosis of psychogenic convergence spasm was made. The patient was then reassured and the symptoms immediately disappeared. She also had psychotherapy and maintained a condition of sustained freedom from symptoms.

Key Words:
Myasthenia gravis, Autoimmune myasthenia, Ocular myasthenia, Anti-AcetylCholine receptor antibodies, Psychogenic convergence spasm, Conversion or hysterical convergence spasm.

Introduction

Psychogenic (hysterical or conversion) Convergence Spasm (PSG) is the most common functional eye movement disorder[1-3]. Understanding of the differential diagnosis, which is broad and includes many organic causes (e.g., stroke), is essential to make an early and accurate diagnosis to prevent complications and initiate appropriate management[3].

We describe a case of a girl with PSG misdiagnosed as ocular myasthenia.

Case Report

A 14-year-old girl was firstly seen by us in the November 2013. She complained of a two-years history of convergent strabismus, diplopia, and reading difficulty due to accommodation problems.

Her family and personal medical history was unremarkable as well as neurological examination except ocular signs. Routine laboratory investigations, brain MRI and multimodal evoked potentials resulted into the normal limits.

In 2011, she received a diagnosis of ocular myasthenia, based on: a) clinical findings; b) Single Fibre Electromyography (SFEMG), which showed a slightly increased jitter; c) the presence of serum antibodies against Acetylcholine-Receptor (Anti-AChR-Ab): 0.96 nanomoles/l – nv: < 0.60 nanomoles/l; and d) the clinical response to Anticholinesterase drugs (oral Pyridostigmine). On the other hand, electromyographic tests of repetitive stimulation and CT scan findings of the thymus were normal.

As the symptoms persisted and significantly impaired girl's studies and social life, she was hospitalized several times. Diagnostic test were also repeated. While neuroradiological study of the mediastinum and repetitive stimulation continued to be normal, single fibre EMG revealed again an increased jitter of 20%. Moreover, anti-AChR-Ab was never longer present and anti-musk antibodies were absent. A course of anticholinesterase drugs (30 to 60 mgs of pyridostigmine every 4 hours) improved strabismus and diplopia, which sometimes disappeared. Unfortunately after few days, the therapeutic effect was no longer present. The diagnosis of ocular myasthenia was then confirmed.

She was also treated with steroids (prednisone 50 mg/day) with rapid benefit, which however was transient and quickly disappeared. In May, July and August 2013 she received 3 cycles of intravenous immunoglobulins: each time: 0.4 g/kg/day for 5 consecutive days. After each treatment the patient had a transient remission but after few days the strabismus, diplopia and the defect of accommodation relapsed. After the last cycle the girl developed an almost complete loss of voice thus suggesting a generalization of myasthenia. Aphonia and later dysphonia reversed within a week.

In November 2013 the patient was referred to us considering our large practice on this topics[4-7].

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A few days before the consultation, aphonia relapsed. To reduce diplopia and to read she was wearing glasses with very thick lenses.

At neurological examination we found: large, normally reagent to the light pupils; marked convergent strabismus not improved by rest and not worsened by repeated contraction. Moreover the patient was aphonic. The remaining neurological examination was within normal limits. Moreover, the patient appeared quite indifferent to her clinical condition. We suspected a functional disorder and we scheduled a second consultation few days later.

At the second consultation, the parents were asked to wait in the waiting room while one of us had a very long conversation with the patient. The girl was invited to seat on the chair of the doctor while the doctor seated on her left so that she could whisper on his ear. Then, the doctor kindly asked if she had had an intense displeasure when she was 10 or 11 old. At the second request, some tears appeared on her eyes and she whispered in the doctor's ear that nobody in previous years had never made this question. Yes, she had had a great dolour when she was 10 and her uncle of 24 years died in a motorcycle accident. This uncle was her best friend and he accompanied her everywhere with his motorcycle. After he died she felt to be completely neglected because her mother was always with her parents who had lost their son.

A female teacher only understood that she had a problem and she was used to talk with this teacher. Unfortunately, the teacher was moved to another school in another city and consequently she remained alone again.

While she was talking, a trickle of voice appeared and the doctor was able to ward off his ear from her lips.

Then, the doctor took for himself a strong candy (with mind and chilli pepper) and offered her one of them. While she was sucking the candy, she said to feel an intense sensation of freshness in the nose and in the throat and the voice further improved.

The doctor said that she was an intelligent and sensitive girl who was suffering for 4 years and the moment had now arrived to enjoy a normal life, or better a joyous life. And she said she desired study to become MD, and to dance and laugh with her friends.

<Now – the doctor added – remove these mole's glasses!>.

She removed the glasses and her eyes were perfectly straight and she had no longer diplopia and could easily read.

When the parents came inside the medical office, everybody was moved: the patient, the parents and the doctor as well.

After this consultation, the patient had no longer muscle and medical problem to date. She had a psychotherapy in her city for six months. Later she wanted to suspend it.

Now she is a 18 young female healthy, she does not assume any therapy, she is finishing the high school with brilliant results and she is preparing the admission test to study medicine.

Discussion

Although it is frequent to misdiagnose myasthenia and treat the patient for other diseases such as stroke, multiple sclerosis, hysteria, eating disorders, etc., it is rare the opposite error, namely to diagnose myasthenia in patients having others disorders[7]. Accommodation problems are very rare in myasthenia while they are common in conversion convergent spasm (transient ocular convergence, miosis and accommodation associated with disconjugate gaze mimicking abducens palsy[1]. In the young patient, in fact, the correct diagnosis was psychogenic (hysterical or conversion) convergence spasm[1-3].

In the light of this, the single detection of circulating AntiAChR-antibodies probably was due to a lab mistake while the increased jitter, found twice at SFMEG, should be considered non-specific at anyway inconsistent. Finally, the positive effect of anticholinesterase drugs and immunosuppressive interventions could be explained with a placebo effect, very common in conversion disorders.

Functional (psychogenic) neurological symptoms are frequently encountered in neurological practice. Cranial movement disorders – affecting the eyes, face, jaw, tongue, or palate – are an under-recognised feature of patients with functional symptoms[2] and can present in isolation or in the context of other functional symptoms. Convergence spasm is the most common functional eye movement disorder[3].

Psychogenic symptoms mimicking a neurological disorder pose an uncomfortable and often frustrating challenge, both in diagnosis and management. Unfortunately, once the diagnosis of a neurological condition is made, it is easily perpetuated without being questioned, which explains the usual diagnostic delay and costs associated with conversion disorders.

Although the specialty of functional neurological disorders has expanded, appreciation of cranial functional movement disorders is still insufficient. Identification of the positive features of cranial functional movement disorders such as convergence might lend diagnostic weight to a suspected functional neurological disorder. Increased understanding of these disorders is also crucial to drive clinical trials and studies of individually tailored therapies[2].

It is important to note that the diagnosis of conversion disorders may be difficult initially for several reasons. In our opinion, physicians are taught often consider and exclude physical disorders as the cause of physical symptoms. Probably, the modern complete division between neurological and psychiatric practice could play a role in this way to proceed. It is the same reason for which it is common experience that some patients with depression are treated by neurologists as having Parkinson disease and some patients with Parkinson are treated by psychiatrists as having depression. Furthermore, physicians are more likely to treat for the more serious condition if they are in doubt of the diagnosis.

Conclusions

When the symptoms do not fulfil diagnostic criteria of a neurological disorder, the objective examination is not neurologically congruous and there is not consistent response to an appropriate treatment, a psychological condition should be actively searched for.

Conflict of interest

The authors declare no conflicts of interest.

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