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CASE REPORT

Three first cases of Meige syndrome in Central Africa

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Abstract: Meige syndrome (MS) is a rare type of focal dystonia characterized by unintentional bilateral activity in facial and mandibular muscles in combination with blepharospasm (spasm in the orbicular muscle). Often unknown cause, MS can be primary (idiopathic) or secondary (to medications or brain degenerative pathologies). Its treatment is based in most cases on botulinum toxin injections, not available in our environment. Nevertheless, clonazepam treatment has allowed us to improve the symptoms of our patients. We report MS in three patients, all with no particular history of consulting for involuntary facial movements. The diagnosis of MS was essentially based on the clinical picture. After treatment with clonazepam, the clinical course in all three patients was marked by the almost complete disappearance of these abnormal movements. In all cases, clinical progress was favorable with an average decline of 12 months; no recidivism was noted.

Keywords: Meige syndrome, abnormal movements, clonazepam, Lubumbashi

1 Introduction

Meige syndrome (MS) is a neurological entity that clinically associates blepharospasm with involuntary movements of the lower facial region and/or masticatory muscles [1]. Different eponyms such as “Wood syndrome”, “Meige syndrome”, “Brueghel syndrome” have been used to describe segmental craniocervical dystonia [2]. It is also called “Brueghel syndrome”, the name of the Flemish painter who is apparently the first to describe this condition (on a painting) in the 16th century. In 2007, Martinez-Castrillo *et al.* [3] suggest that a potter from Moche culture in Ancient Peru (100–700 AC) may be the first artistic representation of blepharospasm with oromandibular dystonia (Meige’s syndrome).The French neurologist Henri Meige described this syndrome in detail in 1910, in its idiopathic form under the name of “median facial spasm”. He also described the extension of dystonia to the pharynx, the jaw muscles, the muscles of the floor of the mouth, the tongue muscles, and the neck muscles. The syndrome is mainly characterized by blepharospasm, oromandibular, and facial dystonia [4]. The term “Meige syndrome” was first proposed by Dr. George Paulson in 1972 for patients with facial muscle spasms, particularly blepharospasm and oromandibular muscle dystonia [5,6]. After several years, Gilbert had used the term “Brueghel syndrome” for a case of oromandibular dystoniawhich differs from MS by the absence of blepharospasm [4]. MS may be primary (idiopathic) or secondary (to drugs or brain degenerative pathologies). Several studies have suggested genetic involvement in the etiology of MS [6,7]. The use of Botox injections is the most effective treatment for MS to date, but its effects are not sustainable. However, most patients are often treated with oral medicines, such as anticholinergics, tetrabenazine (monoamine storage inhibitors), benzodiazepines, and baclofen [6,8]. To our knowledge, these three Congolese cases we report would be the first in Central Africa. This study reports three cases of MS seen in a consulting room in Lubumbashi in the Democratic Republic of the Congo.

2 Case Report

2.1 Case 1

M.L., 49-year-old woman, with no particular history, presented to the Lubumbashi University Clinics in March 2020 for episodes of facial, neck and mouth shakes that affected her speech and diet. The symptomatology would date back to three months of the consultation without

any particular context. No management had been initiated. Given the persistence of these clinical signs, she will decide to consult us for a good management. Her medical and surgical histories were unremarkable, there was no notion of medication (antiparkinsonian, antihistamine or antidepressants).

On neurological examination, there was intense bilateral and rhythmic blepharospasm, divergent strabismus in eyes, photophobia, minor dysphonia and stuttering in the rhythm of contractions, contraction of the facial muscles giving the appearance of a grimace, stuck teeth, a stiff neck with contraction of the neck muscles. These clinical signs were fading for a few seconds and were resuming with increasing intensity. The brain magnetic resonance imaging (MRI) performed was unremarkable. Based on all these elements, we concluded at Meige Syndrome. Therapeutic management was based on pharmacological treatment: the patient received clonazepam at a dose of 4 mg daily in two doses. After two weeks of treatment, complete remission of involuntary movements, dysarthria, and other symptoms was noted. No recidivism was noted after a 10-month decline.

2.2 Case 2

N.K., 55-year-old man, with no particular history, presented to the Lubumbashi University Clinics in January 2021 for involuntary shaking and facial movements, which reportedly started 6 months ago. No medical treatment had been given to him beforehand, but the patient pointed out that he had received medicinal herbs from a traditional practitioner. On neurological examination, we noted a lucid patient with a good mental orientation in space, time, and person; with no language and memory problems. No cranial nerve damage was objectified. Sensitivity to the upper and lower limbs was bilaterally intact. Reflexes were normal and symmetrical at the upper and lower extremities. The assessment of motor function showed facial spasms of excessive and involuntary blinks of the eyelids in both eyes describing blepharospasm associated with spasms in the jaw and neck muscles and dysarthria. Based on these elements, it was highly suspicious that the patient was suffering from Meige syndrome, which is a combination of blepharospasm and oromandibular dystonia. Biological investigations and brain MRI were normal. Clonazepam 4 mg daily twice-dosed treatment was administered to the patient, and the clinical progress was already marked by myorelaxation of the eyelids at one week and at three weeks by a marked improvement in symptoms. No recidivism was noted after a 14-month decline.

2.3 Case 3

F.H., 51-year-old man, with no particular history, had consulted the Lubumbashi University Clinics on June 2020, complained of involuntary movements in the eyelids, mouth, lower jaw and neck. These movements disrupted his diet and his language. The onset of symptoms dated back to two months before his arrival at the hospital. The patient had previously been treated in another medical facility where he had been prescribed methyl-dopa for a month without success. It is the persistence of these involuntary movements that will motivate the patient to consult us.

Neurological examination shows blepharospasms, oromandibular spasms accompanied by contraction of the facial and neck muscles. Dysarthria was also noted. A state of anxiety was objectified in this patient. No conductive or sensorineural hearing loss was found. Examination of other systems was unremarkable. Treatment with clonazepam (4 mg per day in two doses orally) and alprazolam (0.5 mg per day orally) had been administered and showed a marked decrease in these abnormal movements after three weeks of treatment. After a 12-month of follow-up, the patient had no symptoms.

3 Discussion

MS is a rare form of focal dystonia characterized by unintentional bilateral activity in facial and jaw muscles in combination with blepharospasm [9–11]. The onset of the disease usually occurs between 30 and 70 years (the average age is 55.7 years). MS is 2 times more common in women than in men because it has been assumed that specific estrogen receptors make women prone to involuntary muscle spasms [6, 9, 10, 12]. Our patients were 49, 51 and 55 years of age, and therefore were in the most common age group.

MS can be primary (idiopathic) or secondary. Primary forms are the most common. The cause and pathophysiology of primary MS is not clear enough [10]. Since the majority of cases are idiopathic, normal brain imaging is often common. In addition, all patients with possible hereditary or secondary dystonia should undergo brain MRI imaging. The rest of the investigations should focus on suspected etiologies [12]. There are no special investigations to diagnose MS. However, his diagnosis remains based on a complete patient history, external observations and a neurological examination. In our patients, the brain imaging performed (brain MRI) was

unremarkable, which allowed us to orient ourselves much more towards idiopathic MS than secondary MS since there were no other special circumstances in all three cases. Focal dystonia is mainly treated by botulinum toxin injections [9, 10, 13]. Other drug approaches involving anticholinergic agents (such as trihexyphenidyl), benzodiazepines (including clonazepam) and baclofen are commonly used. Recent studies have reported improvement with clozapine and zolpidem [12]. Of all these molecules, clonazepam is the most available and accessible in our environment, which is why we used it as a first-line treatment for our patients. In several cases described in the literature, clonazepam was administered to patients with MS, with improved symptomatology [14, 15]. In a study conducted in Tunisia by Benhouma *et al.* [16] on the clinical and therapeutic aspects of dystonia, clonazepam has been used in a large number of patients with good symptom improvement. This drug has also been tested by Goldberg *et al.* [17], successfully in the treatment of blepharospasm.

4 Conclusion

In the majority of cases, MS is idiopathic; however, careful questioning and clinical examination should guide the clinician so that a reversible condition is not missed. In the absence of the botulinum toxin and baclofen recommended in its management, clonazepam was used successfully in all our three cases.

Data availability

The data used to support the findings of this study are available from the corresponding author (OM) upon request.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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