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Clinical Case

Abnormal Movement and Literature Review on Meige's Syndrome in a Mining Context

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Abstract

We report the case of Meige's syndrome in a 30-year-old young man with no particular history living and working in a mining environment and whose etiological assessment is unremarkable, which classifies him in the vast majority of idiopathic Meige's syndromes until to the contrary. The interest of this case lies in the need to carry out a thorough investigation of all the toxic substances circulating in this environment where mining activity is intensive and to do this, to upgrade the technical platform by calling on collaborations.

Keywords: Meige Syndrome; Abnormal Movements; Dystonia; Blepharospasm; Contracture; Inflammatory; Toxic

Introduction

Meige syndrome is a segmental cranial dystonia characterized by blepharospasm and oromandibular dystonia, often accompanied by abnormal movements of the maxillofacial and neck muscles [1]. It is part of the focal dystonias and is characterized by simultaneous involvement of the upper and lower part of the skull, including binocular spasm of the eyelids (blepharospasm; BSP) and involuntary movements of the jaw muscles (dystonia of the mouth and of the jaw; OMD) [2,3].

The etiology and pathogenesis of this extrapyramidal disorder are unknown. Neurological and ophthalmological examinations often reveal no abnormalities. However, in some cases, infectious, inflammatory, toxic, drug, vascular, tumoral, mechanical, traumatic, genetic causes can be found, but in the vast majority of cases no cause is found.

A few genes are the cause of isolated dystonia, but in the case of combined forms, more than 100 genes have been identified [6]. In all cases, the etiological search is often unsuccessful, but when a cause is found, specific treatment is possible except of course when it is genetic [6].

DYT1, DYT3 and DYT6 dystonias often involve parts of the craniocervical musculature. On the molecular and systemic level of

torsinA, TAF1 and THAP1, respectively, allow us to understand the molecular biology and cellular mechanisms and in particular of craniocervical dystonia [4,7].

Gene expression profiling in rats and functional neuroimaging evidence in humans suggest that DYT1 dystonia is a neurodevelopmental process disorder [4]. At the cellular level, torsin A appears to mediate the interaction between the nuclear envelope and the cytoskeleton [4].

Thus, torsinA mutants can indirectly disrupt the movement of transcription factors and transcripts into and out of the nucleus, respectively. Since TAF1 and THAP1 are transcription factors expressed in the central nervous system, transcriptional dysregulation may be a common underlying pathophysiological mechanism in many forms of primary dystonia.

The sporadic nature of most primary focal dystonias and the imperfect penetrance and variability of some hereditary dystonias raise the possibility that environmental factors play an important role in the pathophysiology of this movement disorder. In all focal dystonias, there may be inappropriate sensorimotor plasticity in peripheral sensorimotor deficits [4].

For example, blepharospasm often begins with symptoms of eye inflammation due to conditions such as dry eye and blepharitis [4]. These patients typically exhibit increased blinking (adaptive response) before developing full-blown blepharospasm (maladaptive response). Similarly, most patients with primary oromandibular dystonia had undergone blunt facial trauma or dental procedures before the onset of movement disorders [4], and in some series more than 20 patients with cervical dystonia One patient reports significant previous trauma to the neck [4]. Based on this clinical information, a rational argument has been made that dystonia is an abnormal sensorimotor integrative disorder [4].

Relatively few pathophysiological studies have focused specifically on head and neck segmental dystonia or in subjects with the blepharospasm plus phenotype [4,5].

Nevertheless, studies of blepharospasm, mastication, and cervical dystonia are clearly relevant to our understanding of segmental dystonia of the head and neck, as focal dystonias share a common genetic and physiological basis. Physiological studies have shown abnormal excitability of interneuron pathways in sensorimotor areas of the brainstem and cerebral cortex in patients with focal dystonia of the head and neck region [4,5].

Brainstem interneuronal pathways have been studied using the blink reflex and the masseterin inhibitory reflex. The recovery cycle of the R2 component of the blink reflex and the SP2 component of the masseter inhibitory reflex is enhanced in patients with craniocervical dystonia. Subjects with isolated cervical dystonia also showed better recovery of R2, suggesting that abnormal interneuronal excitability extends beyond areas of clinical involvement. Dresel., *et al.* [4] used functional MRI with silent events to compare three experimental groups.

Blepharospasm alone, blepharospasm and maxillomandibular dystonia, and controls. Both dystonia groups were shown to have increased somatosensory and caudal activation of the supplemental motor cortex during the whistling task. The shortened cortical telogen phase in the blepharospasm and blepharospasm + oromandibular dystonia experimental groups is consistent with the hypoexcitability of cortical inhibitory neurons in cranial dystonia.

The phenotypic aspect of each focal dystonia is correlated with the age of onset [2]. Blepharospasm and oromandibular dystonia (55.7 years) are more likely to develop 15 years later than other focal dystonias such as writer's cramp (38.4 years) and cervical dystonia (40.8 years) [2]. There is also a gender difference. The female/male ratio is between 1.6.1-3.3:1[2].

The overall prevalence of cranial dystonia and focal dystonia is not well defined. The reported prevalence varies widely between 50 per million for early onset primary dystonia and 30-7320 per million for late onset primary dystonia [8]. A prevalence study in Region A of the Americas estimated it at between 13 and 130 cases per million for blepharospasm and 69 cases per million for OMD [9].

In a multicenter European prevalence study of eight countries, the prevalence of primary focal dystonia in Europe in 2000 was 117 per million, with blepharospasm accounting for 36 per million (95% CI 1-41) [2].

As the disease progresses, involvement of the lower facial and chewing muscles, such as lip pursing, chewing, jaw thrusting, grimacing, jaw opening/closing/clenching, etc., is very common in patients with BSP.

The spasm usually lasts for tens of seconds before spreading to other areas. Because the seizures are less synchronous, the dystonia can last for several minutes. Tonic contractions may be preceded by clonic contractions or hyperactivity.

Clinical observation

He is a 30-year-old man in good health, 5th of 7 siblings from a monogamous family who had a normal primary, secondary and university education, holder of a bachelor's degree in management economics, living and working in a mining town which has had a permanent contracture of the right upper limb for 7 hours in a context of insomnia associated with a mild depressive state treated with Deroxat 20 mg in the evening. The aggravation of the muscular contractures of the right upper limb is the reason for his consultation at the neuropsychiatric center Dr Joseph Guislain in Lumumbaschi.

His background is unremarkable, he does not drink alcohol, he does not smoke or consume psychoactive substances.

The neurological examination is normal apart from abnormal movements made of tonic jerks of the right upper limb associated with contractures of the sternocleidomastoid and of the right trapezius leading to lateroflexion of the neck towards the left with jerk of the head blinking of the eyes and contracture of the corner of the lips on the right side.

Faced with this picture evoking Meige's syndrome and while awaiting the relevant investigations to refine the positive and etiological diagnosis, he is put on Clobazepam, Risperidal and Methylprednizolone respectively at a dose of 2mg every 12 hours, 1mg for 24 hours and 10mg/24H which reduce the frequency and intensity of abnormal movements

Paraclinical assessments

Biological examinations	Value found
NFS	NFS Lymphocytes 45%, Neutrophils 52%, Eosinophils 1% Monocyte0.2% IMG 0.1%, GR 4.24%, HT 33.9%, VGM 79.9% TCMH 29.3%
CRP	6,3g/dl
Urea, Creatinine	15mg/dl 1,4 mg/dl
ALAT	22 U/L
ASAT	32 U/L
Blood sugar	79mg/dl
SRV	Absente
Cupruria, cupremia, ceruloplasmia	Normal except Iron at 187 ug/dl
Antistreptolysine O (ASLO)	Normal
Complete blood ionogram	Normal
CPK and CRP	Normal

Table a

Imaging

MRI performed after the onset of symptoms found no abnormality, particularly in the central gray nuclei.

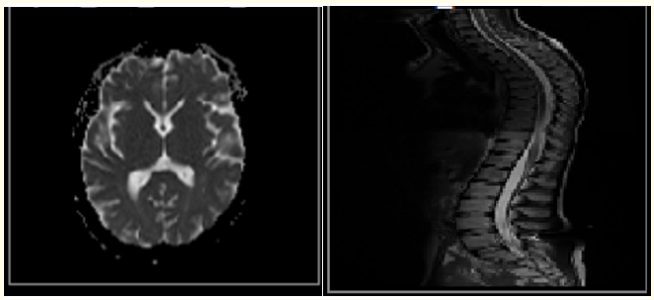

Figure 1,2: Brain and spinal cord MRI Normal.

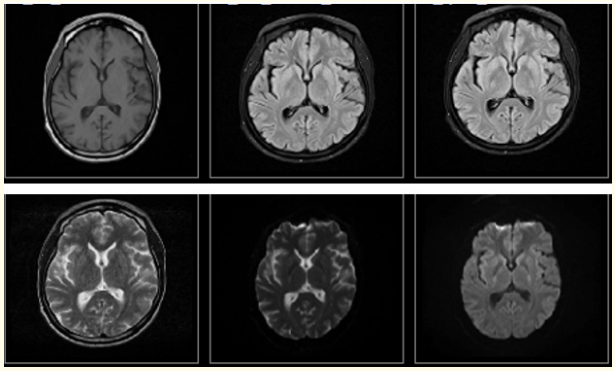

Figure 3: MRI: normal., Electrocardiogram: normal.

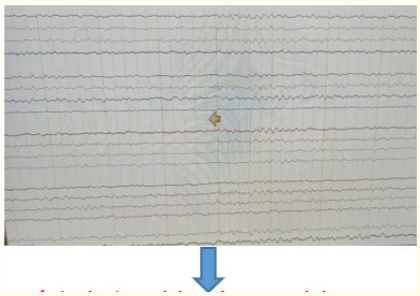

Figure 4: Awakening and sleep electroencephalogram: normal.

Other exams (chest X Ray, Electrocardiogram, Awakeness and sleep Electroencehalogram) : Normal

The evolution was favorable with a regression of the symptomatology and a return home on the 14th day with a normal neurological examination, including the neuropsychological test.

Discussion

In the absence of objectifiable etiological argument in the state of our technical platform, our case must be classified as idiopathic Meige syndromes until proven otherwise. Indeed, the neuroradiological assessment is normal as well as the inflammatory assessment and the search for toxins and responsible drugs. Careful investigation should be considered as soon as the technical means allow it to eliminate the possible role of the numerous toxic substances linked to intensive mining in this region. In addition it should be necessary to perform genetic investigations and functional MRI which are not available in our country.

Conclusion

Our case is similar to the vast majority of idiopathic Meige syndromes. Its originality lies in the mining environment where our patient lives and works. The interest is to underline the need to carry out meticulous investigations to find the toxins which could be responsible and which have not yet been found in this environment particularly exposed by intensive mining. Our situation as a developing country and our very insufficient technical platform constitute a handicap to achieve this.

Bibliography

1. Chang-Geng Song., *et al*. "Successful treatment of the Meige's syndrome with navigated repetitive transcranial magnetic stimulation: A case report". *Brain Stimulation Journal* 15.1 (2022): 96-98.

2. Travis JW Hassell and David Charles. "Treatment of Blepharospasm and Oromandibular Dystonia with Botulinum Toxins". *Toxins* 12 (2020): 0269.

3. Hongying Ma., *et al*. "Syndrome Clinical and Genetic Update".

4. Mark S Le Doux. "Meige Syndrome: What's in a Name?" *Parkinsonism and Related Disorders* 15.7 (2009): 483-189.

5. Hongying Ma., *et al*. "Syndrome: Clinical and Genetic Update".

6. HA Jinnah. "Medical and Surgical Treatments for Dystonia". *Neurologic Clinics* 38.2 (2020): 325-348.

7. H Russmann., *et al*. "Abnormal movements (dystonia, athetosis, chorea, ballism) Movement disorders (dystonia, athetosis, chorea, ballism) ". 1

8. Baptiste Abbar., *et al*. "RET-Rearranged Lung Adenocarcinoma with Paraneoplastic Meige Syndrome". *Journal of Thoracic Oncology* 14.11 (2019): e250-e251.

9. Jiayu Liu., *et al*. "Regional metabolic and network changes in Meige syndrome". *Scientific Reports* 11 (2021): 15753.