Isolated frontalis, corrugator and procerus dystonia – a blepharospasm variant

Distonia isolada dos músculos frontal, corrugador e prócero – uma variante de blefaroespasmo

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Blepharospasm (BSM) is a form of cranial dystonia caused by involuntary bilateral eye closure produced by contractions of orbicularis oculi muscles, often accompanied by dystonic movements of the eyebrows, paranasal, facial, masticatory, labial, lingual, oral, pharyngeal, laryngeal and cervical muscles[1-3]. In most instances, BSM is idiopathic. However, cases associated with lesions in rostral brainstem or other parts of basal ganglia (multiple sclerosis, stroke, thalamotomy and autoimmune disorders) have been described [1,3,4]. BSM is more common in females (3:1 rate) and, in almost three quarters of all cases, after the age of 59 years[1,4].

We report an unusual form of focal dystonia, affecting the frontalis, corrugator and procerus muscles, without any involvement of orbicularis oculi muscles that characterizes BSM.

A 46-year-old Afro-South American male was initially evaluated due to a complaint of mild involuntary forehead muscle movements, leading to significant physical and psychological discomfort. The patient presented the symptoms since childhood, and the intensity of the contractions worsened until the age of 35 years, remaining relatively stable since then. Neurological examination showed symmetrical involuntary movements of frontalis, corrugator and procerus muscles, completely sparing the orbicularis oculi muscles. Additionally, he presented upward oculogyric deviations (Figure). The movement disorder disappeared when his eyes were closed and returned immediately after opening them. There were historic or clinical data suggestive of a tic disorder. Laboratory tests and cranial CT and MRI scans were unremarkable. Initially, he was treated with flufenazine, haloperidol and clonazepam with no clinical effect, as a differential diagnosis of tic disorder was made.

Our next approach was for intramuscular botulinum toxin type A injections. After informed consent was obtained, a total dose of 100/IU botulinum toxin type A (Dysport, Ipsen, UK) was injected in the frontalis, corrugator and procerus muscles. On a three-week follow-up, the dystonic symptoms and signs were completely abolished.

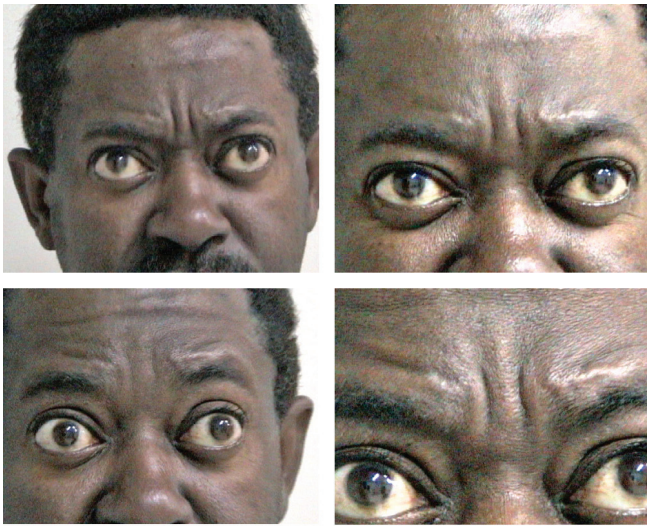

Figure. The patient showed abnormal and involuntary movements of frontalis, corrugator and procerus muscles, bilaterally, without any compromising of orbicularis oculi muscles. Also, he presented oculogyric deviations (up), during the dystonic movements.

BSM is relatively uncommon in men, especially before the age of 50 years. Its hallmark is the involvement of the orbicularis oculi muscles that is essential for the diagnosis. This partially explains the difficulties we found in regards to the differential diagnosis of the movement in this patient as he presented the symptoms since childhood. The main differential diagnosis of our case was simple chronic motor tic although was no evident premonitory phenomenon. The abnormal movements were neither intermittent, repetitive nor abrupt. Additionally, there was no supressibility and response to the traditional treatment.

Nonetheless these unusual features, we assumed the possibility of an unusual presentation of cranial dystonia with sole bilaterally involvement of frontalis, corrugator and procerus muscles. We believe that the upward eye deviations were a sensory trick that transiently relieved the involuntary movements.

We also highlight the fact that this unusual presentation of dystonia is almost indistinguishable from essential BSM, except by the non-involvement of the orbicularis oculi muscles.

To best of our knowledge, this is the first report of sole frontalis, corrugator and procerus dystonia. Recently, Hirota et al.[5] reported two cases of dystonic frowning. The first patient presented involuntary frowning and grimacing as the sole symptom, with no eyelid involvement. The second had typical symptoms of BSM only in the early phase of the disease, later the pattern of contractions changed and involuntary frowning and grimacing predominated. As most focal dystonias, these cases also presented excellent response to the use of botulinum toxin type A.

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