Journal of the Neurological Sciences 372 (2017) 162–170

Contents lists available at ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns

Review Article

Meige's syndrome: History, epidemiology, clinical features, pathogenesis and treatment

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ARTICLE INFO

Article history:
Received 14 September 2016
Received in revised form 31 October 2016
Accepted 21 November 2016
Available online xxxx

Keywords:
Blepharospasm
Dystonia
Botulinum toxin
Deep brain stimulation

ABSTRACT

'Meige's syndrome' is a type of cranial dystonia characterized by blepharospasm and oromandibular dystonia and can be associated with complex movement of lower facial muscles, mouth, jaw, tongue, pharyngeal and cervical muscles. Frequently, blepharospasm is the earliest clinical manifestation, which spreads over a period of time to involve other cranial and extra-cranial muscles. Common characteristics of this syndrome are well known, but their variety is wide. Different eponyms such as "Breughel syndrome", "Wood syndrome", "Blepharospasm plus", "Segmental cranial dystonia" and "Segmental cranio-cervical dystonia" have been used to describe this entity with numerous anatomical variations. In the majority of the patients Meige's syndrome is primary or idiopathic, where the cause of spasm is not known, however secondary cases can occur following prolonged use of neuroleptics or secondary to underlying brain disorders. This syndrome has also been described in patients with essential tremor, Parkinson's disease and atypical Parkinsonism. Neurophysiological features are similar to other focal dystonia characterized by abnormal plasticity and impaired inhibition. Most of the patients are successfully treated with injection of botulinum toxin, however deep brain stimulation has emerged as a good therapeutic option in intractable patients.

The objective of this review is to understand whether patients who develop Meige's syndrome are different from patients who manifest blepharospasm or oromandibular dystonia alone.

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http://dx.doi.org/10.1016/j.jns.2016.11.053
0022-510X/

1. Introduction

In 1910 Henry Meige, a French neurologist described a disorder characterized by dystonic spasm involving majority of the facial muscles [1–3]. Since Meige's original description, Altrocci, Paulson, Marsden and Jankovic described similar syndrome characterized by blepharospasm and oromandibular dystonia [4–7].The neurologists have often described this condition as "Meige's syndrome" and "Meige syndrome" which is confusing, as 'Meigs' syndrome has also been used to define the triad of benign ovarian tumor, hydrothorax and ascites [8,9]. We will be using the term Meige's syndrome in this review, which is based on the PubMed search with an aim to focus on the historical controversy regarding the different nomenclatures, epidemiology, clinical features, pathophysiology and current treatment. We have also included patient's videos from our database to show the clinical features and anatomical variations.

2. Search strategy

Relevant studies on "Meige's syndrome" were reviewed using the PubMed search till 18th April 2016. Additionally, we also searched for 'Blepharospasm' as significantly large number of studies on this topic, have also included 'Meige's syndrome' patients. A total of 377 articles on 'Meige's syndrome', 337 articles on 'Meige syndrome', 1839 articles on 'Blepharospasm', 34 articles on 'spread of blepharospasm', 39 articles on 'segmental cranial dystonia', 18 articles on 'segmental craniocervical dystonia', 122 articles on 'pathophysiology of Meige's syndrome', 466 articles on 'pathophysiology of Blepharospasm', 5 articles on 'neuroimaging in Meige's syndrome', 33 articles on 'neuroimaging in blepharospasm', 104 articles on 'Botulinum toxin in Meige's syndrome', 769 articles on 'Botulinum toxin in blepharospasm', 27 articles on 'Deep brain stimulation in Meige's syndrome' and 27 articles on 'Deep brain stimulation in blepharospasm' were found. The final reference list was based on the relevance to the topic of review.

3. History

Dr. Horatio Wood, a Philadelphia neurologist, first described blepharospasm and other cranial dystonias in 1887 (Fig. 1) [10]. He believed that the lower facial spasm was a counteractive measure to relieve the eyelid spasms. However, the syndrome was named after Dr. Henri Meige, a French neurologist who in 1910 described 10 patients with bilateral facial convulsions in the vicinity of the midline, designating it "spasm facial median" [1]. He also observed similar clinical characteristics in muscles other than facial region, which included pharynx, jaw, floor of the mouth and tongue. Henri Meige was not sure about the pathogenesis and he hypothesized that since facial convulsions are suppressed by will and they disappear during sleep they were more likely to be psychogenic, but he also believed that spasms beyond the facial region may be due to the disturbances provoked by an irritative lesion in the bulbopontine region. In 1925, Henderson described 135 patients of essential blepharospasm and postulated that facial spasms are due to the organic changes in the basal ganglia [11] >40 years later in 1972, George Paulson described 3 patients with blepharospasm and oromandibular dystonia as Meige's syndrome [5]. Subsequently, in 1976 Marsden described 39 patients with blepharospasm and oromandibular dystonia, but, he named this as Brueghel's syndrome thanking Dr. R. E. Kelly, who pointed out that Pieter Brueghel, the Elder recognized this syndrome [6]. Two decades later in 1996 Gilbert published a case of jaw opening dystonia only (without blepharospasm) as Brueghel's syndrome and emphasized that this syndrome must be differentiated from Meige's syndrome, where patients have blepharospasm and oromandibular dystonias [12]. He pointed out that occasionally both conditions may occur in the same patient, but overall Meige's syndrome was far common than Brueghel's syndrome [12]. In 1983, Jankovic and Ford described 100 cases of blepharospasm and orofacial-cervical dystonia as "Meige's syndrome" [7]. In 1994 Casala and Deuschl discussed different types of focal dystonia of cranial nerve innervated muscles; blepharospasm, mandibular dystonia, spasmodic dystonia, lingual dystonia, pharyngeal dystonia, external laryngeal

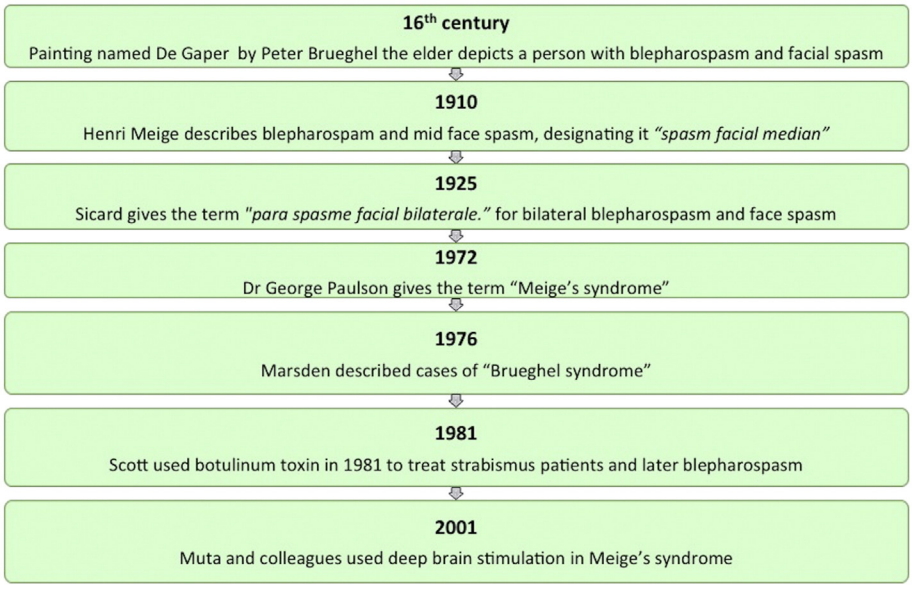

Fig. 1. History of Meige's syndrome.