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Meige syndrome: What's in a name?

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ABSTRACT

Frequently, blepharospasm is associated with involuntary movements of the platysma, lower face and masticatory muscles. Similarly, masticatory dystonia may occur in isolation or in combination with dystonia of other cranial and cervical muscles. The non-possessive and possessive forms of Meige and Brueghel syndromes have been variably and imprecisely ascribed to various anatomical variations of craniocervical dystonia. Herein, the origin of eponymic terms as applied to craniocervical dystonia is reviewed as support for proposed elimination of these eponyms from clinical usage. Although the term "segmental craniocervical dystonia" more accurately captures the combination of blepharospasm and dystonia of other head and neck muscles, delineation of craniocervical subphenotypes is essential for etiological/genetic and treatment studies. To conclude, the clinical features, epidemiology, pathophysiology and therapeutic management of segmental craniocervical dystonia are examined with a particular focus on "blepharospasm-plus" subphenotypes.

Contents

1. Introduction

The terms "Meige's syndrome" and "Meige syndrome" are often used by neurologists and other clinicians to describe the combination of blepharospasm and involuntary movements of the lower facial and/or masticatory (jaw) muscles. Application of "Meige's syndrome" and other eponyms to the various forms of dystonia is problematic for a multitude of reasons. First of all, Meige, a physician, did not suffer from the syndrome that bears his name. Along this line, the possessive form of eponyms has been discouraged by the Council of Science Editors [1] and the father of Online Mendelian Inheritance in Man®, the late Dr. Victor McKusick [2]. Second, Meige was not the first person to describe the combination of blepharospasm and dystonia of other cranial muscles [3]. Lastly, "Meige's" or "Meige syndrome" could be confused with Meigs syndrome which is defined as the triad of a benign ovarian tumor, ascites and hydrothorax [4].

2. Historical perspective

Dr. Horatio Wood, a Philadelphia neurologist, first drew attention to blepharospasm and other cranial dystonias in 1887 [3].

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Wood briefly mentioned facial and oromandibular dystonia in his textbook on disorders of the nervous system [3]. He stated, "The contraction is tonic, causing a complete closure of the eye, and consequent blindness. This is accompanied by innumerable bizarre grimaces, due to the efforts of the antagonistic muscles to overcome the force which is closing the lids". Clearly, his second sentence was incorrect.

In 1910, Henri Meige, a French neurologist, described approximately ten patients with involuntary closure of the eyelids [5]. Blepharospasm was associated with involuntary contractions of the jaw muscles in only one of these patients. Over 60 years later, an American neurologist, George Paulson, reported three patients with blepharospasm and oromandibular dystonia and emphasized the probability of a common pathophysiological basis [6].

In 1976, David Marsden, an English neurologist based in London, called attention to a work of art, *De Gaper*, by Pieter Brueg(h)el the Elder, a Flemish Northern Renaissance painter, in an article on blepharospasm and oromandibular dystonia [7]. Brueg(h)el the Elder was not a physician and his painting of a yawning subject has nothing to do with dystonia. Pieter Brueghel the Elder dropped the 'h' from his name in 1559, one year after painting the *De Gaper* [8]. Brueghel the Younger was also a painter, further confounding historically exact usage of this eponym [8].

Careful inspection of the relevant medical literature over the past 30 years indicates that "Brueghel" and "Meige" syndromes remain poorly delineated. One author, Dr. Gordon Gilbert, suggested that the essential sign of "Brueghel syndrome" is "a widely and dystonically opened jaw [9]". In reality, however, jaw-opening dystonia may occur in the setting of segmental, multifocal or generalized dystonia and may be associated with blepharospasm [10–12]; application of "Brueughel syndrome" to these cases would be unnecessarily complicated and confusing. Marsden, in the title of his 1976 article on the subject, actually defined blepharospasm–oromandibular dystonia syndrome as "Brueghel's syndrome" and he did, in fact, use the possessive eponym [7].

The eponymic terms "Meige's syndrome" and "Meige syndrome" are used much more frequently than the 4 variations of "Brueghel syndrome". PubMed (www.ncbi.nlm.nih.gov) search (April 18, 2009) with the terms "Brueghel's syndrome," "Bruegel's syndrome," "Brueghel syndrome," and "Bruegel syndrome" generated 9, 0, 5, and 0 hits, respectively. In contrast, PubMed search with the terms "Meige syndrome" and "Meige's syndrome" generated 263 and 95 hits, respectively. For comparison, "craniocervical dystonia," "craniocervical dystonia" and "cranial dystonia" produced 6, 29 and 77 hits, respectively. Even in recent years, application of the eponymic terms remains highly variable. "Meige syndrome" and "Meige's syndrome" have been ascribed to both primary and secondary dystonias and diverse craniocervical anatomical patterns. For instance, Zesiewicz and colleagues reported substantial improvement in "Meige's syndrome" with levetiracetam treatment although their patient also exhibited cervical dystonia [13]. For an even more divergent example, a 15-year-old boy with "Meige syndrome" was described as showing "a stiff face, labial incompetence, prominent mental creases and dimples with a lot of involuntary blinking and involuntary movements of the lower face [14]". Kraft and Lang [15] defined "Meige's syndrome" as blepharospasm associated with dystonic movements of other muscle groups in the face, neck or limbs. Clearly, the time has come to terminate usage of poorly-defined eponymic terms for dystonia of the craniocervical region.

3. Anatomically-based classification of dystonia by distribution

In 1984, an *Ad Hoc* Committee of the Dystonia Medical Research Foundation developed a widely-accepted definition of dystonia and classification of dystonic movements [16]. Dystonia was defined as a motor syndrome characterized by sustained muscle contractions, usually producing twisting and repetitive movements or abnormal postures. It was noted that dystonia is often precipitated by action and almost all dystonic movements share a directional quality that is typically sustained, sometimes for only an instant. The *Ad Hoc* committee classified dystonia by age at onset, etiology and distribution [16].

Classification by distribution includes the following categories: focal dystonia, segmental dystonia, generalized dystonia, multifocal dystonia and hemidystonia [16]. The term "focal dystonia" indicates involvement of a single body part. Common names such as blepharospasm, spasmodic dysphonia, writer's cramp and spasmodic torticollis are often assigned to the focal dystonias. The term "segmental dystonia" denotes involvement of two or more contiguous regions of the body. The segmental dystonias are subdivided into regional categories: cranial, axial, brachial and crural [16]. Within this classification scheme, segmental "cranial" dystonia indicates involvement of any combination of musculature in the head and neck region. In reality, however, the neck and mandible are not parts of the cranium [17]. Therefore, the combination of blepharospasm, masticatory dystonia and cervical dystonia, for example, is more precisely classified as segmental "craniocervical" dystonia rather than segmental "cranial" dystonia [18]. Alternative terms such as "facio-cervical" are less precise since the masticatory muscles neither arise nor attach to the face. In fact, most facial muscles attach to the cranium and/or mandible. The term "segmental craniocervical" encompasses all muscle groups in this body segment with "cranio" serving as the rostral component of this segment and cervical forming the caudal portion.

The diagnosis and anatomical classification of dystonia remains a clinical exercise profoundly influenced by experience and training. Regional involvement may be subtle, intermittent and task-specific [19]. Moreover, the origins and insertions of many muscles are located in different anatomical regions of the body (e.g., levator scapulae, digastric, platysma, semispinalis capitis and longissimus capitis). Accordingly, clear demarcation of "focal" from "segmental" dystonia may be difficult in many patients. For instance, cervical dystonia patients with retrocollis often exhibit mild, electromyographically-demonstrable involvement of the upper thoracic paraspinous musculature.

There are several reasons why use of the term "segmental craniocervical dystonia" may be more accurate than "segmental cranial dystonia" in most patients with "blepharospasm-plus" subphenotypes. First, involuntary contractions of the platysma muscle can be seen in an important percentage of patients with blepharospasm. In these subjects, mild contractions of the platysma muscles may be time-locked with those of the orbicularis oculi muscles. The platysma is innervated by the facial nerve, the same nerve which innervates the orbicularis oculi muscles in the eyelids [17]. The platysma is a broad, thin, superficial muscle that extends from the upper chest, shoulder and clavicle upwards to the chin, mandible and lower face [17]. Second, the pharyngeal musculature, which can be affected in subjects with blepharospasm, encompasses both the cranial and cervical regions [17,20]. Similarly, several muscles involved in jaw-opening originate in the cervical region [17,21]. Based on these considerations, "segmental craniocervical dystonia" is an anatomically more precise term that envelops virtually all past clinical utilization of the terms "Meige syndrome" and "Breughel syndrome".

The anatomical complexity of the craniocervical region also confounds employment of currently-available rating scales for dystonia [22–24]. The widely-used Burke-Fahn-Marsden (BFM) rating scale includes the following regions: eyes, mouth, speech and swallow, neck, right arm, left arm, trunk, right leg and left leg.