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Dystonia with Tremors: A Clinical Approach

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1. Introduction

Dystonia commonly accompanies tremors but the prevalence of the association is controversial. Oppenheim had already described that tremors were associated with dystonic symptoms in "Dystonia Musculorum Deformans" in the early 20th century (Oppenheim et al, 1911, as cited in Jedynak et al., 1991). Yanagisawa analyzed idiopathic dystonia with electromyography and found that dystonia was associated with rhythmic activity in all of the patients (Yanagisawa & Goto, 1971). In a genetic and clinical population study on dystonia, 80% of the population had tremors for generalized dystonia (Larsson and Sjogren, 1966). Marsden reported that 14% of patients with generalized nonfamilial idiopathic dystonia presented with tremors (Marsden, 1974). In addition, 68% of patients with cervical dystonia had head tremors (Pal et al., 2000). However, Rondot examined 132 patients with cervical dystonia, which revealed rhythmic activity and upper limb tremors in 40% and 21% of the patients, respectively (Rondot et al., 1981, as cited in Jedynak et al., 1991). In a survey on writer`s cramp, hand tremors were reported in almost half of the subjects (Sheehy, 1982). In addition, Jankovic investigated 350 patients diagnosed with Essential tremor (ET), based on the presence of tremors in the head, hand, or voice in the absence of any other diseases that may cause tremors. Forty-seven percent of these subjects also had dystonia (Lou and Jankovic, 1991). Therefore, the prevalence of dystonia with tremors varies greatly depending on the reports.

Dystonic tremor syndrome has been under-recognized and sometimes mistaken as ET or even Parkinson`s disease (Elble and Deuschl, 2011). However, dystonic tremor syndrome is not just ET but also a distinct clinical entity, and has the possibility of having a secondary cause (Bain, 2009; Cho et al., 2000; Jankovic & Linden, 1988; Kim & Lee, 2007; Oyama et al, 2011; Schneider et al., 2007; Vidailhet et al., 1998; Yoon et al., 2009). Moreover, the progress and treatment of dystonic tremors are different from other tremor disorders (Gironell & Kulisevsky, 2009).

However, dystonic tremor syndrome is still under debate and different definitions have been proposed (Deuschl et al., 1998).

This chapter will focus on the clinical criteria and differential characteristics of dystonic tremor syndrome.

2. Clinical criteria of dystonic tremor syndrome: According to the involved site

Dystonic tremor is a relatively new classification of tremor. The Movement Disorder Society (MDS) proposed a consensus statement for the tremor in 1998 (Deuschl et al, 1998).

According to these criteria, dystonic tremor syndromes were divided into three types: dystonic tremor, tremor associated with dystonia, and dystonia gene-associated tremor.

2.1 Dystonic tremor

Dystonic tremor means tremor in a body part affected by dystonia. That is to say, the tremor and dystonia occur simultaneously in the same body part such as the arm or neck. This is usually a focal, postural, or kinetic tremor but usually not seen during complete rest (Deuschl et al, 1998). Typical examples of this type are a dystonic head tremor, which is a head tremor in patients with cervical dystonia, and a dystonic writing tremor, which is a writing tremor in patients with writer`s cramp.

2.2 Tremor associated with dystonia

This tremor occurs in a body part not affected by dystonia, but the patient has dystonia elsewhere (Deuschl et al, 1998). It is uncertain whether this type of tremor is the cormorbid occurrence of ET along with dystonia (Lou & Jankovic, 1991) or is a distinct entity (Deuschl et al., 1997; Munchanu., 2001; Shaikh et al., 2008). A typical type is an upper limb postural tremor in patients with cervical dystonia.

2.3 Dystonia gene-associated tremor

This type of tremor is an isolated finding in patients with a dystonia pedigree. A typical example of this type is an isolated tremor occurring in a patient with first-degree relatives with spasmodic torticollis (Deuschl et al, 1997; Yanagisawa et al., 1972).

2.4 Variability in the definition of dystonic tremor

Quinn reported that in the absence of any alternative causes for their tremor, dystonic tremor and tremor associated with dystonia should be called dystonic tremor (Quinn et al., 2011). The prevalence and other clinical details of dystonic tremor are variously reported since the clinical criteria of dystonic tremor are not clearly defined. This chapter describes dystonic tremor syndrome following the MDS criteria.

3. Differential characteristics of dystonic tremor syndrome

The dystonic tremor is significantly different from disorders with pure tremors. In addition, the tremor associated with dystonia has also been reported recently to be different from other pure forms of tremors combined with dystonia. However, the clinical significance of the dystonia gene-associated tremor is not known.

3.1 Dystonic tremor

In a study on idiopathic dystonia with electromyography, Yanagisawa described that dystonia was stimulated by postural effort, and that, largely irregular, sometimes regular, tremulous muscle activity was observed during a dystonic posture (Yanagisawa & Goto, 1971). In a study on dystonic tremors with electromyography, the dystonic tremor was shown to be postural, localized, and irregular in amplitude and periodicity; and absent during muscle relaxation, exacerbated by smooth muscle contraction, and associated

frequently with myoclonus (Jedynak et al., 1991). The frequency of the dystonic tremor is mostly below 7Hz, and very rarely, rest tremors may occur (Deuschl, 1998, 2001). The dystonic tremor may have some specific features of dystonia such as "geste antagoniste" (sensory trick) (Jahanshahi, 2000).

3.2 Tremor associated with dystonia

Tremors of the hands can be seen often in patients with cervical dystonia (Couch, 1976). There were some controversies whether this type of tremor is the same as ET or not. ET and cervical dystonia may be physiologically and possibly also genetically related. Cervical dystonia has been reported in 0.6~30% of patients with ET (Critchley, 1972; Baxter and Lal, 1979; Martinelli and Gabellini, 1982; Rajput et al., 1984; Lou and Jankovic, 1991; Koller et al., 1994; Tallon-Barranco et al.,1997, as cited in Munchau et al., 2001). Additionally, postural and kinetic tremors are found in 4-55% of patients with cervical dystonia (Patterson and Little, 1943; Couch, 1976; Chan et al., 1991; Lang et al., 1992; Dubinski et al., 1993; Deuschl et al., 1997, 1998, as cited in Munchau et al., 2001). In 1991, Lou and Jankovic reported 47% of patients with ET had dystonia, but this analysis found no support for the differentiation of ET subtypes although it was heterogenous in its clinical presentation (Lou and Jankovic, 1991).

However, in a study on tremors with 55 cervical dystonia patients, hand tremors in patients with cervical dystonia more closely resembled an enhanced physiological tremor than a dystonic tremor or ET (Deuschl et al 1997). In addition, arm tremors in patients with cervical dystonia was found to develop either before or simultaneously with the onset of torticollis; such a temporal relationship does not correspond to a dystonic tremor either (Munchanu et al., 2001). Besides, the temporal relationship and physiological quantity is also different. The irregularity of the tremor was significantly greater (~50%) in hand tremors associated with cervical dystonia than that of ET (Shaikh et al., 2008). Moreover, the latency of the second agonist EMG burst was later in ET than in CD patients during ballistic wrist flexion movement (Munchau et al., 2001). These findings suggest that the mechanism for the tremor associated with dystonia may differ from that of ET.

3.3 Dystonia gene-associated tremor

This type of tremor was reported in a large pedigree of "Dystonia Musculorum Deformans" of Japanese descent with autosomal dominant inheritance (Yanagisawa et al., 1972).

4. The clinical approach to dystonic tremors

It is difficult to discriminate a dystonic tremor from ET and myoclonic dystonia and from psychogenic and Parkinsonian tremors. There are some observations that help to differentiate these features.

4.1 Dystonic tremor versus Essential tremor

As mentioned above, a dystonic tremor has an irregular, broader range of frequency than that of ET. Myoclonus sometimes can present in a dystonic tremor, but it is never seen in ET. The dystonic tremor is more localized and less symmetric, that is, it occurs in one arm and hand (Yanagisawa & Goto, 1971).

There may be diagnostic ambiguity in cases of head tremors only. How can this type of tremor be differentiated? A dystonic head tremor has a sensory trick (Deuschl et al., 1992). The occurrence of the sensory trick is useful in the differential diagnosis of a head tremor because the sensory trick is found in as many as 90% of the patients with cervical dystonia but not in patients with ET (Jahanshahi, 2000; Elble & Deuschl, 2011). In addition, the dystonic head tremor appears in large amplitude when the affected body part is placed in a position opposite to the major direction of pulling by the dystonia, but the tremor disappears or decreases when the body part is positioned where the dystonia wants to place it (Fahn, 2009). Moreover, cervical dystonia can have hypertrophy of the affected muscles (Jankovic, 2007) and 75% of patients with cervical dystonia have neck pain (Chan et al., 1991) but never in ET.

Less regular,
Asymmetric
Myoclonic component
Sensory tricks
Aggravation for specific posture or null point
Muscle hypertrophy
Pain

Table 1. Clinical features indicative of a dystonic tremor in an isolated head tremor

4.2 Dystonic tremor versus myoclonic tremor

A dystonic tremor has rhythmic activity and appears when a posture is assumed. However, in myoclonic dystonia, a burst of muscular activity can be recorded even at rest although it is facilitated by postures and movements, and the burst of muscular activity can recur at irregular intervals. Myoclonus can present in addition to the dystonic tremor (Jedynak et al, 1991). However, if myoclonus occurs consecutively, it is difficult to draw a line between a dystonic tremor and myoclonic dystonia (Jedynak et al, 1991).

4.3 Dystonic tremor versus psychogenic tremor

A psychogenic tremor can be confused with a dystonic tremor since the duration of the tremor burst in the dystonic tremor is widely variable reflecting its jerky nature and is similar to some psychogenic tremors (McAuley and Rothwell, 2004). However, the psychogenic tremor has psychogenic signs, multiple somatizations, secondary gain, or is related to an injury or event (Elble, 2000).

4.4 Dystonic tremor versus Parkinsonian tremor

A dystonic tremor may present with Parkinsonism, which can lead to a misdiagnosis of PD. SWEDD (Scan Without Evidence of Dopaminergic Deficit) means there can be cases in which some patients with Parkinson-like tremors have no dopaminergic deficit and therefore, do not have Parkinson`s disease (Schwingenschuh et al., 2010). Adult-onset dystonic tremor is one of the causes of SWEDD (Schneider et al., 2007). However, the two conditions are distinguishable by the presence of a jerky tremor, head tremor, dystonic voice, rapid emergence of a postural tremor, normal olfaction, lack of response to dopaminergic medication, relatively stable natural history, and no progression towards

developing features other than the tremor and dystonia suggesting a dystonic tremor rather than PD (Schneider et al., 2007; Bain, 2009).

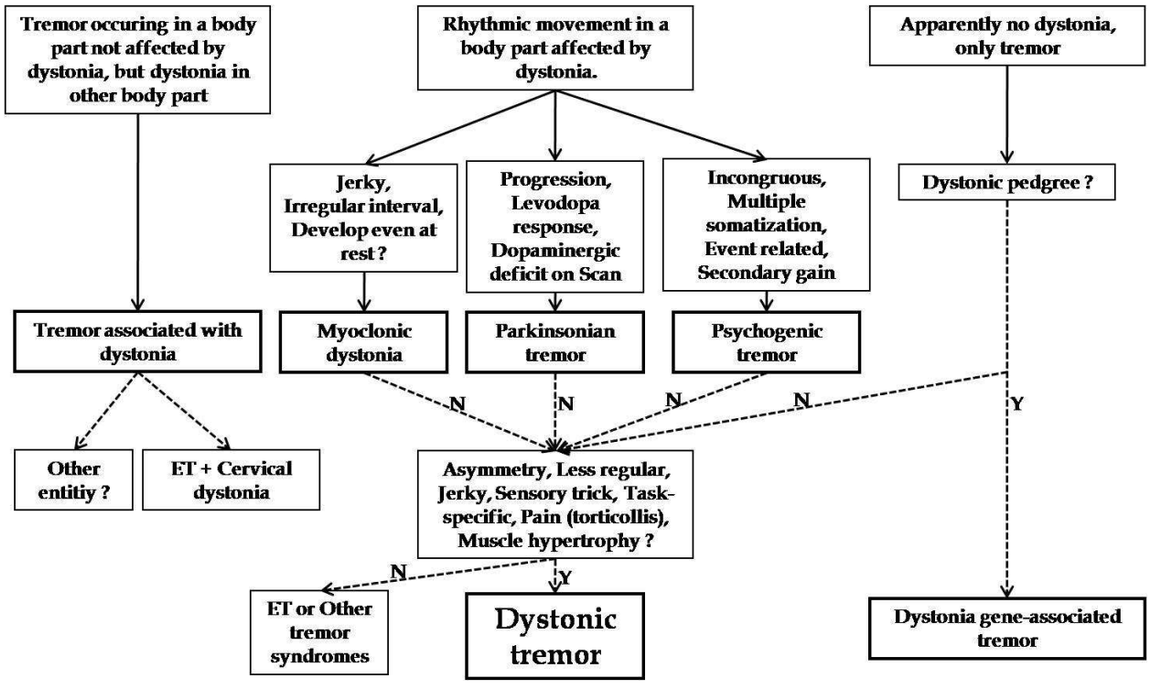

Fig. 1. Clinical approach to dystonic tremor syndrome (Y: yes, N: no)

5. The etiology of dystonic tremor syndrome

Typical primary dystonic tremors are dystonic head tremors and hand tremors in patients with writer`s cramp. However, there are many non-primary causes of dystonic tremor syndrome including Parkinsonism, SWEDD, Wilson disease, NBIA (neurodegeneration in brain iron accumulation), and peripheral trauma and following brain lesions such as thalamus and parietal lesions (Bain, 2009; Cho et al., 2000; Jankovic & Linden, 1988; Kim & Lee, 2007; Oyama et al, 2011; Schneider et al., 2007; Vidailhet et al., 1998; Yoon et al., 2009).

6. The mechanism of dystonic tremor syndrome

The underlying mechanism of dystonic tremor syndrome is not well known. Hallet has proposed that the sensory tricks in dystonic tremors are related to the basic mechanisms underlying the dystonia rather than being a specific feature of the dystonic tremor (Hallet, 1995). Moreover, one widespread notion is that it may be related to the mechanism of dystonia most likely generated within the basal ganglia loop (Deuschl & Bergman., 2002). However, dystonic tremors may also be caused by peripheral mechanisms (Jankovic & Linden, 1988).

7. Conclusion

Dystonic tremor syndrome is distinct clinical entity. Knowing the clinical characteristics of dystonic tremor syndrome is important to help discriminate it from other tremor disorders and to manage it.

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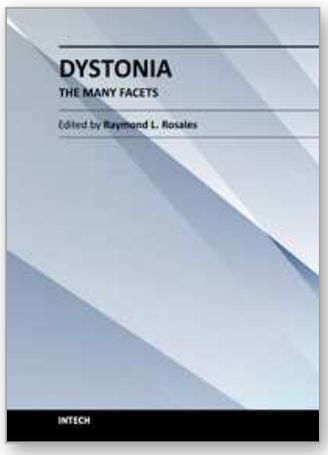

Dystonia - The Many Facets

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Dystonia has many facets, and among those, this book commences with the increasingly associated genes identified, including a construct on how biology interacts with the dystonia genesis. The clinical phenomenology of dystonia as approached in the book is interesting because, not only were the cervical, oromandibular/lingual/laryngeal, task-specific and secondary dystonias dealt with individually, but that the associated features such as parkinsonism, tremors and spasticity were also separately presented. Advances in dystonia management followed, and they ranged from dopaminergic therapy, chemodenervation, surgical approaches and rehabilitation, effectively complementing the approach in dystonia at the clinics. A timely critical pathophysiologic review, including the muscle spindle involvement in dystonia, is highlighted at the book's end.

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