Other movement disorders

Michele TM Hu

Abstract

Abnormal movement disorders are common in neurology but frequently challenge even the most experienced movement disorder neurologist. Many patients defy rigorous categorization, with the specific challenge of interpreting the patient whose movements look odd, but who just might have an organic movement disorder. This chapter will cover movement disorders other than those related to the parkinsonian syndromes. Emphasis will be given to those movement disorders more commonly encountered by the general physician and general practitioner, with illustrative case presentations seen by the author in a district general hospital setting. Tremor, dystonia, chorea, myoclonus, tic disorders and drug-induced movement disorders will be covered with emphasis on clinical presentation, correct diagnosis and subsequent management.

Keywords chorea; drug-induced movement disorders; dystonia; myoclonus; tics; tremor

Tremor disorders

Functional brain imaging in the form of single photon emission computed tomography (SPECT) and ^{18}F-dopa positron emission tomography (PET) scans allow measurements of nigrostriatal dopamine, the chemical that is deficient in idiopathic Parkinson's disease (PD). Symptom onset in PD is estimated to occur at a mean putamen ^{18}F-dopa uptake of 75% of normal levels.[1] These techniques have revealed that 15% of patients in whom a neurological diagnosis of PD has been made on clinical grounds have no evidence of dopaminergic deficit on functional scans.[2] What is the correct alternative diagnosis in this fascinating subgroup of patients who have scans without evidence of dopaminergic deficit or 'SWEDDS'? Clinically, these patients often present at a similar age to PD patients (mean age 63 years) with an asymmetric, resting arm tremor and reduced arm swing. In contrast to PD patients, however, head tremor is relatively common, and they frequently have dystonia with task specificity (see next section) in the tremulous limb, leading to the correct diagnosis of dystonic tremor. Furthermore, these patients do not have true akinesia with progressive fatiguing or decrement of alternating movements that is so characteristic of PD.[2] Dystonic tremor may be an under-recognized cause of late-onset tremor and in fact more common than essential tremor. The typical patient with essential tremor often recalls a barely perceptible hand tremor from their early 20s to 30s but seeks medical attention at a later age. The tremor affects both arms and tends to be more symmetrical than dystonic tremor. Head involvement is common, and the tremor is markedly reduced after 1–2 units of alcohol. As in dystonic tremor, a positive family history is encountered in approximately one-third of essential tremor cases.

Treatment of non-parkinsonian tremor

Correct diagnosis of a tremor disorder is essential as it dictates appropriate treatment. Dystonic tremor may be worsened by levodopa[1] and is more appropriately treated with clonazepam, an anticholinergic or botulinum toxin injections to the dystonic limb. Essential tremor is best treated with a beta-blocker or primidone, although the latter is often poorly tolerated due to systemic side effects.

Dystonia

Dystonia is probably the least recognized and poorest understood of the common movement disorders. More than any other kind of movement disorder, patients with dystonia are not uncommonly referred to psychiatric colleagues. Although prevalence studies are limited, dystonia is probably half as common as idiopathic PD. Fahn's description of dystonia as 'a syndrome dominated by sustained muscle contractions, frequently causing twisting and repetitive movements or abnormal postures' provides a workable definition of dystonia.[3]

Dystonia has many classifications according to the age of onset, the aetiology or the part of the body affected. General dystonia affects the whole body, hemidystonia (more likely with structural brain lesions) affects the whole of one side of the body, and focal dystonia is denoted by its focus (e.g. torticollis in the neck, writer's cramp in the dominant hand or blepharospasm in the eyelids). Focal dystonia tends to develop later in life than most childhood-onset forms of generalized dystonia. Tremor in association with dystonia is not uncommon (see above). Often, the abnormality becomes apparent only with movement and is referred to as 'action dystonia'. If the abnormality occurs only on performance of a particular action, such as writing or playing a musical instrument, it is said to be 'task specific'.

Treatment of dystonia

Focal dystonia is best treated with botulinum toxin injections into the overactive muscles. This avoids the systemic side effects common with oral medications such as anticholinergics, benzodiazepines and tetrabenazine, which are moderately effective for dystonia. An example of a patient with focal neck dystonia (torticollis) is discussed in Table 1. Photographs before and after treatment with botulinum toxin are shown in Figure 1. Patients with severe, generalized dystonia may need a combination of anticholinergic, tetrabenazine and dopamine blockade. Deep brain surgery, in the form of pallidal stimulation, can produce excellent benefit in selected patients.

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Case history 1

This woman first noticed an involuntary head tilt to the right at the age of 27 years. Symptoms progressed rapidly over the next year, with associated neck pain and increasingly prominent head tremor. She described the effective use of a sensory trick or geste, where touching her chin gently with her fingers enabled her to keep her head comparatively straight. Botulinum toxin injections into predominantly right-sided neck muscles produced an excellent benefit to her pain and head posture that has been maintained over 8 years, with injection intervals of 3 months (see Figure 1).

Table 1

Chorea

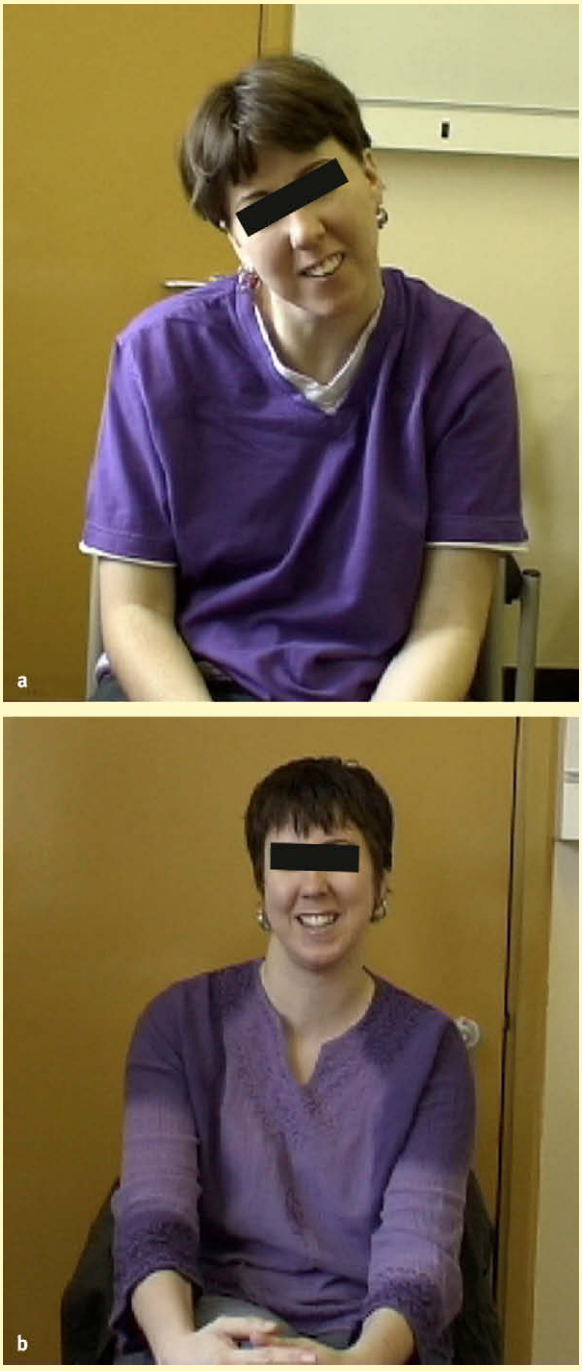
The term 'chorea' derives from the Greek for 'the art of dancing' and as such is an accurate description for these sinuous, fidgety, involuntary movements. The causes of chorea are multiple and include Huntington's disease, pregnancy (chorea gravidarum), drugs (including oral contraceptive pill), streptococcal infection (Sydenham's chorea), systemic lupus erythematosus, antiphospholipid syndrome, vascular chorea, senile chorea and neuroacanthocytosis. Several electrolyte and hormonal abnormalities have been associated with chorea, including, most commonly, hyperthyroidism, but also hypo- and hypernatraemia, hypocalcaemia, hypomagnesaemia, hypoglycaemia and hypoparathyroidism. An example of a patient presenting with chorea in the context of a psychotic disorder is discussed in Table 2.

Treatment of chorea

This largely depends on establishing the correct diagnosis of the cause of chorea. Many patients are not disabled by their chorea, and therefore treatment is inappropriate. Patients with disabling chorea, particularly if associated with psychotic features, as in the case described above, or patients with Huntington's chorea may benefit from dopamine receptor antagonists, such as olanzapine, sulpiride, quetiapine or risperidone. Common side effects include parkinsonism, akathisia or tardive dystonia. Dopamine-depleting agents such as tetrabenazine can be effective but often cause depression that can be difficult to treat. Low doses of a long-acting benzodiazepine, such as clonazepam, can be effective without inducing major side effects.

Myoclonus

Myoclonus is defined as shock-like involuntary movements arising from the central nervous system.[5] Positive myoclonus arises from brief bursts of muscle activity; less commonly, negative myoclonus arises from a sudden, short inhibition of ongoing muscle activity. Myoclonus can be classified according to its site of origin (cortical, brainstem and spinal). Cortical myoclonus tends to be most marked in the distal limb. If widespread, myoclonus is multifocal, sometimes elicited by movement, local touch or visual stimuli. An example of this is post-anoxic myoclonus (Lance and Adams syndrome), commonly seen following respiratory hypoxic events. Cortical myoclonus can be related to



Photograph of patient with focal cervical dystonia (torticollis) **a** before and **b** 3 months following botulinum toxin injections to neck muscles.

Figure 1

neurodegenerative disorders such as Huntington's disease, prion disorders, Alzheimer's dementia, or mitochondrial or coeliac-related encephalopathy. Table 3 is an example of cortical myoclonus encountered in a district general hospital setting. Brainstem myoclonus leads to generalized myoclonic jerks that affect proximal muscles more than distal. An exaggerated startle response, particularly to auditory stimuli, is common. Spinal myoclonus is