Facial movement and expression disorders

Part 2. Excessive movement

The face displays a wide range of involuntary movements that originate at all levels of the motor system. In Part 2 of this article, excessive facial movement is discussed, complementing the discussion of facial weakness in last month's issue of Medicine Today.

JASON WARREN
BMedSc, MB BS(Hons)
CHRISTOPHER KNEEBONE
MB BS, FRACP

Dr Warren is Registrar, National Hospital for Neurology and Neurosurgery, London, UK; Dr Kneebone is Senior Visiting Neurologist, Department of Neurology, Royal Adelaide Hospital, Adelaide, SA.

Disorders with excessive facial movement can be identified from the pattern of muscular activity they produce (see Table 1). The common theme in many of these disorders is loss of control over dopaminergic pathways that normally regulate involuntary facial expressions.

For an overview of neuroanatomy for classifying and understanding disorders with excessive facial movement, refer to the box entitled 'How are facial movements and expressions controlled?' in Part 1 of this article.

Tics

Tics are irregular, brief, stereotyped movements that preferentially affect the face and upper body. They may be triggered by a variety of emotional and physical stimuli (such as anxiety, anger or fatigue) and are characteristically abolished by others (such as intense absorption, alcohol or febrile illnesses). Unlike most movement disorders, which are aggravated by stressful mental activity, tics can be suppressed by concentration and reappear with relaxation. Production of the tic is accompanied by a sense of release of inner tension.

The most common examples are single, stereotyped twitches of isolated muscles (simple tics). More complex movements and gestures, such as blinking, sniffing, grinding the teeth or shrugging also occur, and may be difficult to distinguish from the facial mannerisms universal in the general population.

Tourette's syndrome is a dominantly inherited condition in which multiple tics develop that affect several muscle groups. Onset is usually between the ages of 5 and 15 years, and the distribution of tics alters over time. Involuntary vocalisations are

IN SUMMARY

- **The common theme in many disorders of excessive facial movement is loss of control over the dopaminergic pathways that regulate involuntary facial expression.**
- **Tics probably result from loss of regulatory control normally exerted by the cerebral cortex over dopaminergic and other neurotransmitter systems, and may be suppressed by dopaminergic antagonists.**
- **Brief muscular twitches and jerks are observed in facial myoclonus, which may appear after an hypoxic brain injury such as cardiac arrest or other encephalopathy, and should be distinguished from focal motor seizures involving the face.**
- **Benign essential tremor may produce rhythmic oscillations of the head (titubation), jaw or voice. Propranolol can be used if there are no contraindications.**
- **In hemifacial spasm and focal dystonias, injection of affected muscles with botulinum toxin type A (Botox) is a safe and now first line therapy.**

continued

Table 1. Excessive facial movement: diagnosis and management

Features	Site of lesion	Possible diagnoses	Investigations	Management
Repetitive twitching, sequential muscle activation				
Associated ipsilateral limb muscle involvement, secondary generalisation	Cortex	Focal seizures	Brain CT or MRI, EEG	Anticonvulsant, referral
Irregular brief stereotyped movement suppressed by concentration				
Associated upper body involvement, vocalisations	Cortex, subcortical structures	Tic (simple, Tourette's syndrome or other rare cause)	–	Referral, ?dopamine antagonist
Irregular writhing, flowing movement				
Associated generalised movement	Basal ganglia	Generalised chorea (Huntington's disease, other rare causes)	Genetic testing	Referral
Associated orolingual movement, neuroleptic use	Basal ganglia	Tardive dyskinesia	–	Stop neuroleptic use
Sustained abnormal contraction				
No associated features	Basal ganglia, brainstem	Focal or generalised dystonia (idiopathic, Wilson's disease, rare secondary forms)	Brain MRI (or copper studies for generalised dystonia)	Referral (or botulinum toxin for focal dystonia)
Intermittent or twitchy, quivering contractions of one eyelid				
Signs of brainstem disturbance or no associated features	Uncertain	Myokymia of the orbicularis oculi	–	Reassurance
Intermittent or twitchy quivering contractions involving muscles on one side of the face				
No associated features	Pons	Facial myokymia		Referral, ?carbamazepine, ?steroids
Rhythmic oscillation of head, jaw, voice				
Associated hand tremor	?Brainstem	Benign essential tremor		Propranolol trial, referral
Brief jerks				
Associated cardiac arrest, hypoxia	Brainstem	Myoclonus		Referral
One brow lifts, eye closes, angle of mouth twitches repetitively				
No associated features	Facial nerve	Hemifacial spasm		Referral, botulinum toxin

a characteristic feature; however, these are usually grunts rather than intelligible obscenities. Stereotyped, obsessional and ritualistic behaviours may also occur.

Tics probably result from loss of regulatory control exerted by the cerebral cortex over dopaminergic and other neurotransmitter systems projecting from subcortical structures. Tics may be suppressed by dopaminergic antagonists.

Chorea

Chorea that affects the face may be part of a generalised disorder such as Huntington's disease, an autosomal dominant condition that also produces an early onset dementia that can be confirmed on genetic testing. Chorea may also be seen in elderly edentulous patients but the mechanism in these cases is not known.

Dyskinesia

Tardive dyskinesia

A variety of tardive facial movements may occur after chronic exposure to dopamine receptor antagonists such as neuroleptics (all classes) and antiemetics (metoclopramide and prochlorperazine). The sinuous, writhing, flowing orolingual movements of tardive dyskinesia are very characteristic, and include lip smacking, tongue protrusion, licking, chewing movements and vocalisations. Older patients and women are at greatest risk from tardive dyskinesia which may increase in severity or persist after cessation of neuroleptics.

Sustained, abnormal patterns of muscle contraction (tardive dystonia) and repetitive, purposeless, stereotyped movements (tardive tics and stereotypies) often coexist with orofacial chorea. Fast chittering movements of the lips and jaw have been described vividly as 'rabbit syndrome'. Observation off medication is the recommended treatment. It appears that newer generation antipsychotics such as risperidone (Risperdal) and olanzapine (Zyprexa) carry a lower risk of development of tardive dyskinesia.

Drug-induced dyskinesia

Levodopa and dopamine agonists may produce orofacial choreiform movements, especially in the context of advanced Parkinson's disease. Supersensitive dopamine receptors at the site of action in the basal ganglia are probably the common factor in these drug-induced conditions.

Drug-induced facial movement disorders and examples of causative agents are listed in Table 2.

Dystonia

Dystonias are characterised by abnormal, sustained patterns of muscular contraction. Yawning may be regarded as a physiological form of dystonic facial grimacing, and a variety of pathological dystonias affect the facial musculature.

Focal dystonia

Blepharospasm (abnormal, involuntary bilateral contraction of orbicularis oculi) is a common example of a focal dystonia – eye closure initially takes the form of frequent blinking but may progress to functional blindness in severe persistent cases. Injection of affected muscles with botulinum toxin type A (Botox), which blocks acetylcholine release at the neuromuscular junction and produces a local paralysis lasting several months, is a safe and now first line therapy for blepharospasm (Figure 1).

The lower face is involved in oromandibular dystonia, which takes a number of forms (unilateral or bilateral, with sustained jaw closure or opening, in combination with blepharospasm or alone).

Generalised dystonia

Facial dystonias may also be part of a more generalised disorder. Most cases are idiopathic. Drugs are an important cause (Table 2), and acute dystonic reactions to metoclopramide and neuroleptics are easily recognised.

Numerous other conditions occasionally produce symptomatic dystonias,

Table 2. Facial movement disorders: associated drugs

Tics and stereotypies
- Amphetamines, methylphenidate
- Antipsychotic neuroleptics
- Dopamine agonists
- Levodopa

Chorea
- Amphetamines, methylphenidate
- Anticholinergics, tricyclic antidepressants
- Anticonvulsant toxicity (carbamazepine, phenytoin)
- Antihistamines, cimetidine
- Cocaine, xanthines
- Digoxin toxicity
- Lithium toxicity
- Oral contraceptives

Tardive dyskinesia
- Antiemetics (metoclopramide, prochlorperazine)
- Antipsychotic neuroleptics (all classes), including:
  - Phenothiazines (e.g. chlorpromazine, trifluoperazine, pericyazine)
  - Butyrophenones (haloperidol)
  - Thioxanthenes (thiothixene)
  - Pimozide

Drug-induced dyskinesia
- Dopamine agonists (bromocriptine, pergolide)
- Levodopa

Parkinsonism
- Antiemetics
- Antipsychotic neuroleptics
- Lithium
- Methyldopa
- Tetrabenazine

Dystonia
- Antiemetics
- Antipsychotic neuroleptics
- Lithium

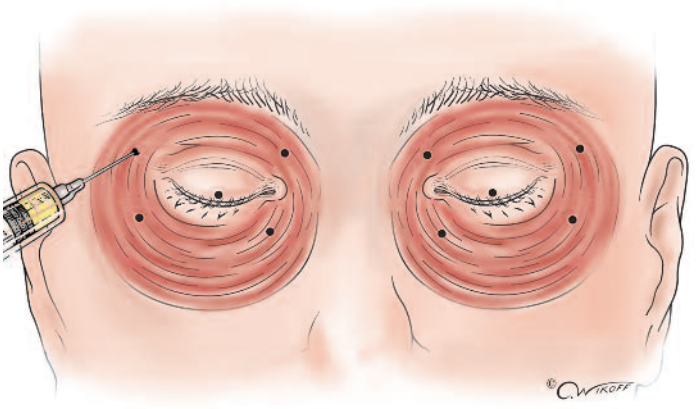

Figure 1. Injection of the orbicularis oculi muscle with botulinum toxin type A is used to treat blepharospasm (bilateral) and hemifacial spasm (unilateral). The needle enters the muscle obliquely to avoid injecting too deeply.

including:

- focal hemisphere lesions
- various neurodegenerative disorders (notably Wilson's disease, a rare but treatable disorder of copper metabolism)
- upper brainstem strokes
- multiple sclerosis.

Classic examples, which are now seen infrequently, include masseteric spasm (trismus) accompanying tetanus and quinsy, and the 'sneering grin' (risus sardonicus) of tetanus.

Facial myokymia

Facial myokymia is characterised by localised, quivering muscle contractions, which occur most commonly in the orbicularis oculi muscles of tired or anxious people. Myokymia involving multiple muscles on one side of the face is a relatively rare phenomenon that results from irritation of facial nerve fibres within the brainstem, as might occur with an acute demyelinating plaque of multiple sclerosis and with infiltrative lesions such as pontine glioma.

Facial myoclonus and focal motor seizures

Brief muscular twitches and jerks are observed in facial myoclonus, which may appear after an hypoxic brain injury such as cardiac arrest or other encephalopathy. These movements should be distinguished from focal motor seizures involving the face – the latter may show a sequential spread of activity or 'march' over a number of seconds, involving ipsilateral limb muscles or leading to a generalised convulsion.

Benign essential tremor

Benign essential tremor may produce rhythmic oscillations of the head (titubation), jaw or voice. There is usually an accompanying tremor of the hands, which responds to a small dose of alcohol in approximately 50% of cases, and there may be a history of similarly affected relatives.

Propranolol ([Deralin, Inderal], commencing with 40 mg twice daily) can be used if there are no contraindications; however, head tremor often responds less well than hand tremor. It should be noted that prominent titubation is rarely due to Parkinson's disease.

Hemifacial spasm

Hemifacial spasm is characterised by bursts of twitching that typically begin around the eye and spread to other muscles on the same side of the face. It is more common in women, and usually appears in the fourth or fifth decade of life. Twitches are typically spontaneous but may increase with stress, fatigue or voluntary facial movements.

Although the origin of hemifacial spasm remains controversial, in many cases there is good evidence of a pulsating, ectatic vessel loop exerting pressure on the facial nerve. This may give rise to ectopic impulses in the nerve where peripheral myelin joins central myelin, as this zone is relatively more excitable.

Botulinum toxin type A is safe and now first line therapy in hemifacial spasm (Figure 1). Microvascular decompression of the facial nerve via a posterior craniotomy in selected patients is a definitive procedure that avoids the need for repeated injections.

The choice of injection site in hemifacial spasm exemplifies the wider social impact of involuntary facial movements. In Anglo-Saxon communities, the involuntary wink is much disliked for its prurient connotations, and the patient's preference is generally for injection of orbicularis oculi, sparing the expressive angle of the mouth.

Conclusion

The diagnosis of disorders involving excessive facial movement can usually be made on the basis of the history and examination findings. Occasional patients will need further investigation to seek underlying pathology.

Hopefully, the introduction of newer neuroleptic agents and the avoidance of long term use of antiemetics (which are often prescribed as treatment for dizziness) will see a decline in the frequency of tardive dyskinesia.

The use of botulinum toxin has been a significant advance in the treatment of blepharospasm and hemifacial spasm. MT

Last month, Part 1 of this article discussed disorders that result in unilateral and bilateral facial weakness.