PostScript

LETTERS

Tourette syndrome and dystonia

The coexistence of tics and dystonia in the same patient,[1] and occurring individually in different members of the same family,[2] has been reported in the literature. This has raised the question as to whether patients with Tourette syndrome (TS) could be at a higher risk for dystonia, and if there is a common pathophysiological mechanism for the expression of tics and dystonia.[2] We sought to determine the prevalence of dystonia among patients with TS using a large international database.

Methods

Using the Tourette Syndrome International Database Consortium (TIC), which is comprised of patients with TS seen by neurologists and psychiatrists worldwide, we searched for cases with a coexisting diagnosis of dystonia. The database was established in 1992 with the aim of learning more about the variability in clinical samples of TS among sites. In all, 28 countries are represented in the consortium, with 52 participating sites. Of the clinicians participating in the consortium, 71% are psychiatrists, 23% are neurologists, 4% are pediatricians and 2% are medical geneticists. At the time of analysis, the database contained 6654 patients, with an average age of 15 years when first seen and entered into the database (range 1–77), and a male predominance of 82%. In all, 695 patients (10%) aged 30 years at the time of registration were in the database. Information collected on the TIC data entry form include the medical specialty of the clinician entering data, family history of TS and related comorbidities, peak tic severity, drugs for tics, perinatal problems, and other neurological, psychiatric or medical diagnoses or problems. Unlisted neurological, psychiatric and medical problems are entered into a text field. There is no specific question addressing the presence of dystonia in the data entry form.

Results

Nine cases of dystonia were found in the database, giving a prevalence rate of 1352 per 1 000 000. Additional history was available from eight of the nine cases. These eight cases came from two clinicians, a neurologist (J Jankovic) and a neuropsychiatrist (K Muller-Vahl), both specialised in movement disorders. In one case, the dystonic hand postures had resolved on follow-up examination 2 years later, making a diagnosis of dystonic tics more probable. In three cases with dystonic hand postures noted on examination, there was a history of significant perinatal complications. In one case, the patient had drug-induced cervical dystonia. The remaining three cases—one with dystonic hand postures, one with left foot dystonia and one with new onset blepharospasm—had no attributable secondary cause.

Discussion

Population studies of idiopathic focal and generalised dystonia reveal an estimated prevalence of 330 per 1 000 000.[3] Since probably only three of our nine cases are idiopathic, this would give a prevalence rate of 451 per 1 000 000 in our sample, which is quite similar to this estimate. More recent epidemiologic assessments of the prevalence of primary dystonia have been divided into early-onset and late-onset cases, and have assessed more ethnically homogenous samples. For example, the prevalence estimate for early-onset dystonia in Ashkenazi Jews from New York is 111 per million, and 600 per million for late-onset dystonia in northern England.[4]

Population rates of secondary dystonia are more difficult to find. The only published figure we were able to find was a prevalence rate of 10 700 per million in the general community aged 50–89 years.[5] The rate of secondary dystonia in the TIC database was 902 per million. It is difficult to make comparisons however, since our clinical sample consists mainly of children in whom one might expect a lower rate of secondary dystonia than in the population of older people.

In addition to clonic tics, the majority of patients with TS also have dystonic tics. Dystonic tics differ from clonic tics in that they are comparatively slow, with sustained twisting or pulling movements, which produce temporarily maintained abnormal postures. Dystonic tics can present diagnostic difficulties to the clinician, but can be distinguished from idiopathic dystonia by history. Diagnostic uncertainty regarding the nature of these movements could lead to both under- and over-representation of dystonia cases in the database.

It is possible, since the data entry form did not specifically ask about the presence of dystonia, that cases were missed and the prevalence rate of dystonia in our TS sample is therefore underestimated. This possibility could further be supported by the fact that the majority of physicians participating in the consortium are psychiatrists rather than movement disorders neurologists. Although more subtle forms of dystonia could not be easily recognised in patients with TS by physicians not specialising in movement disorders, missed cases were probably mild and did not cause significant disability. The clustering of dystonia cases in two clinics specialised in movement disorders could support the possibility of under-recognition in the total survey population but could just as easily suggest a referral bias of atypical cases to these centres, particularly since no other case of concurrent tics and dystonia were reported from other participating movement disorders specialty clinics. The results of this study give an important first estimate of the prevalence of dystonia in patients with TS.

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Crossed aphasia during migraine aura: transcallosal spreading depression?

Transient neurological symptoms in full awareness allow us to understand functional brain organisation at first hand. Indeed, descriptions provided by patients or their families are usually accepted as adequate evidence to localise the side of brain dysfunction during transient ischaemic attacks and partial epilepsies.

Language is lateralised to the left cerebral hemisphere in most right-handed individuals, and crossed aphasia (that is, a language disorder secondary to a right hemisphere lesion in a dextral pertson) is a rarity (1–4%) in otherwise healthy subjects.

Over the past 10 years, as a result of my interest in headache and behavioural neurology, I have observed five patients with crossed aphasia during migraine auras. Patients were examined in an outpatient clinic, and systematically recorded. None of them had any factor that could account for an atypical dominance for language—that is, all patients were right handed, with no personal or family history of left-handedness, previous brain injury, epilepsy, developmental language disorders or learning disabilities. All had normal imaging examinations (MRI of brain in three and CT scan in two subjects) that excluded a structural lesion. Patients were contacted by telephone in 2005–6. *The International Classification of Headache Disorders*, second edition (ICHD-II) diagnosis criteria[1] were checked, based on the patients' account of their usual attacks. New

Table 1 Patients: previous headache history and relevant clinical data

n	Age (years), sex	Handedness Oldfield index	Age at migraine onset (years)	Usual migraine attacks			Attacks with crossed aphasia					Imaging
				MWA	VARS	MwoA	Duration of aura	Aphasic symptoms	Localising signs	Attack witnessed	Repetition of CA attacks	
1	19, M	RH 100	18	Visual, sensory aphasic aura	8	No	20–90 min	Anomia	L hemianopia L paresthesias L hemiparesis	Yes	Yes	CT MRI
2	46, M	RH	36	Visual and aphasic aura	10	No	10 min	Anomia semantic paraphasias	L hemianopia	Yes family	Yes	CT
3	20, F	RH 100	9	Visual and sensory aura	8	Yes	60 min	Anomia speech programming impairment	L hemianopia L paresthesias	No	Yes	CT
4	23, M	RH 80	12	Visual aura	8	No	4 h	Cannot organise sentences paraphasias	L hemianopia L paresthesias L hemiparesis	Yes ER	No	CT MRI
5	34, M	RH 100	12	Visual sensory aura	5	Yes	60–120 min	Stereotype ''hum'' paraphasias	L paresthesias	Yes ER	Yes	CT MRI

CA, crossed aphasia; ER, emergency room; F, female; L, left; M, male; RH, right handed; MWA, migraine with aura; MWoA, migraine without aura; VARS, Visual Aura Rating Scale.

episodes of aphasic auras were investigated, visual aura symptoms were evaluated by the Visual Aura Rating Scale[2] and right-handedness confirmed by the Edinburgh Handedness Inventory.[3] Table 1 summarises the clinical data.

All patients described typical aphasic symptoms (disorders of expressive language, with speech reduced to a stereotype, paraphasias, neologisms, word finding or syntactic difficulties) preceded by left-sided, unilateral, visual, sensory or motor symptoms that localised the dysfunction primarily to the right hemisphere. Patients either had one or more witnesses to the attack (including emergency room physicians who observed them during the attacks), who could confirm language impairment and symptom localisation, or had repeated and stereotyped attacks, providing consistency about the lateralisation of the aura symptoms.

Although all patients fulfilled ICHD-II criteria for migraine (with or without aura), from the account of their usual attacks, in three cases the episode with crossed aphasia was beyond the expected attack duration or included motor symptoms, suggesting sporadic hemiplegic migraine. However, in all subjects, aphasic symptoms built up in the sequence of unilateral progressive typical visual or sensory phenomena and reverted completely within 24 h, making the diagnosis of migraine more likely than stroke. Patients with visual auras scored >5 points on the Visual Aura Rating Scale, which predicts the diagnosis of migraine, and those with sensory symptoms had cheiro-oral or brachiolingual topography, which is also typical of migraine.

Diagnosis of crossed aphasia usually requires the demonstration of a unilateral lesion of the right cerebral hemisphere—that is, it precludes its diagnosis in transient neurological dysfunctions, when patients are often observed after the end of the attack and structural lesions may not be found. This limits its diagnosis in migraine aura, but should not prevent it, because there seems to be no reason to reject a type of evidence (report of symptoms, personally or by observers) that is the basis of clinical diagnosis of other conditions (transient ishaemic attacks, for instance).

Assuming that the phenomenon underlying the aura is unilateral, these five cases suggest that crossed aphasia can be diagnosed on clinical grounds during migraine auras. That assumption is now possible, as functional brain imaging performed during the aura has shown that the underlying phenomenon is primarily neurogenic, unilateral, starts at the visual cortex and spreads slowly forward, corresponding to the timing of clinical symptoms.[4 5] The unilateral nature of aura has also been emphasised by the ICHD-II,[1] which requires the symptoms to be unilateral.

The interest of these cases is that they clearly show that aphasia can occur in association with right hemisphere dysfunction during migraine aura. This observation may unveil pathogenic mechanisms involving the aura or the effect of repeated neuronal dysfunction in language organisation.

Atypical language organisation in migraine has some theoretical support by analogy with other pathologies. Cerebral organisation for language is rather plastic, as language may develop in the right cerebral hemisphere in several conditions (early left hemisphere lesions, arteriovenous malformations and, particularly, in epilepsy, an independent factor responsible for language shift, even when the epileptic focus is localised outside the classic language areas, possibly by causing repeated transient dysfunctions of left hemisphere activity). Migraine is known to cause repeated episodes of transient neuronal dysfunction, especially during the aura, and this factor could stimulate atypical forms of neural organisation. This question is relevant to functional imaging studies, given the high prevalence of migraine in the adult population. It can now be clarified by functional imaging, which was not possible to perform in these patients.

An alternative explanation for crossed aphasia aura is the possibility that the phenomenon underlying the aura travels between the two cerebral hemispheres. Migraine aura has been attributed to cortical spreading depression, a phenomenon first described in rodents, where it has been observed to unfold to the other hemisphere through transcallosal fibres. To date, transcallosal spreading has not been observed in migraine. Although functional images of the aura describe the phenomenon as unilateral, or much more pronounced on the symptomatic side, most reports include few patients with typical auras, which does not cover all possible variations of this phenomenon. If cortical spreading depression also moves transcallosally in humans, then crossed aphasia should not be diagnosed in this specific context.

Both hypotheses may be correct, and we cannot exclude the possibility that this group of patients is heterogeneous and includes patients with atypical dominance (those with stereotyped attacks) and cases with atypical spreading (prolonged attacks).

''Bedside to bench'' research is necessary and justified to understand possible variations of cortical spreading depression and language dominance in migraine.

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