A case of acute chorea

An 18 year old female was admitted with an acute onset of a movement disorder.

HISTORY

The patient presented with worsening 'spasms' of her limbs. They had come on gradually three days prior to presentation and appeared worse in the right hand. She had no voluntary control over the movements, which consisted of twisting motions of the limbs with some more jerky and violent movements superimposed. She also had some involuntary and rather jerky head turning movements. She felt that her speech was affected, in that she was slurring her words and reported difficulty controlling her tongue. Whilst walking she was 'thrown off-balance' by some of the movements. Of note, she had had cut her hand several days before the onset of the illness with associated erythema and evidence of infection of the palm of the right hand. She had not had a sore throat.

Previous medical problems had included a deep vein thrombosis at the age of 14. Investigations at this time had revealed positive lupus anticoagulant in her blood, and raised anti-cardiolipin antibodies (IgG 26 (normal: 0-20); IgM 97 (normal: 0-20)). Thus a diagnosis of anti-phospholipid syndrome had been made. She was commenced on warfarin at that stage, but due to poor compliance, she had discontinued the medication 2 years ago. On her current admission she was on depot contraceptive but no other medications and denied any recreational drug usage. She had a family history of stroke on her mother's side of the family. She was a smoker.

EXAMINATION

On examination she was restless with generalized chorea. This was most marked on the right hand side of the body and she had prominent episodes of head-turning. Her walking was abnormal with severe choreiform intrusions into her gait. She had a mild dysarthria, but no other neurological signs. Her general and cardiological examination was normal.

INVESTIGATIONS

The chorea was investigated with the following tests:

- routine haematological, metabolic and endocrine tests were normal
- coppers and caeruloplasmin levels normal
- MRI scans of the brain on two occasions were normal
- again she had a positive lupus anticoagulant and raised anti-cardiolipin antibodies (IgG 19 (normal: 0-20); IgM >100 (normal: 0-20))
- Streptococcal serology on admission revealed a raised Streptolysin O titre 400 (normal: 0-200) and raised anti-DNAse 980 (normal: upto 85). One week later her Streptolysin O titre remained rasied at 400 and her anti-DNAse levels were further elevated at >1440, indicating recent Streptococcal infection
- an assay for anti-basal ganglia antibodies (ABGAs) was performed at the Institute of Neurology, and these were reported as being positive

DIAGNOSIS

Acute chorea has been associated with anti-phospholipid syndrome and also as a post infectious sequela of Streptococcal infection (Sydenham's chorea). The patient fulfilled criteria for both potential diagnoses. ABGAs are commonly found in cases of Sydenham's chorea (Church et al, 2002).

TREATMENT

In view of the persistence of lupus anti-coagulant and anti-cardiolipin antibodies, she was anti-coagulated again on the advice of the haematologists. She was also commenced on Penicillin V to cover Streptococcal infections. Her chorea responded reasonably well to sulpiride. Follow-up two weeks later revealed marked improvement of her chorea.

REFERENCES

Cervera R, Asherson RA, Font J *et al* (1997) *Chorea in the anti-phospholipid sydrome* Medicine **76(3)**: 203-212

Church AJ, Cardoso F, Dale RC *et al* (2002) *Anti-basal ganglia antibodies in acute and persistent Sydenham's chorea* Neurology **59**: 227-231

Alastair Wilkins
is ACNR's Case Study Co-ordinator. He is Specialist Registrar in Neurology in East Anglia, having trained in Cambridge, Sheffield and London, and has just finished a PhD investigating potential mechanisms of axon loss in multiple sclerosis.

Choreiform movements were most marked on the right, with head-turning movements

Look out for the article on chorea later this year, which will appear as part of our Management Topic on movement disorders in ACNR magazine.

Correspondence address:
Alastair Wilkins, Addenbrooke's Hospital, Cambridge.
E-Mail. aw255@cam.ac.uk