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. 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 and raised anti-cardiolipin antibodies (IgG 19 (normal: 0-20); IgM >100 (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 raised 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

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.