3. Hughes' syndrome and its various guises

In your own general practice, do you have any patients with two or more miscarriages? Or with migraine, or transient ischaemic attacks or mild strokes possibly occurring at a young age?

Do you have patients with atypical multiple sclerosis, or suspected Alzheimer's disease? What about cases of deep vein thrombosis (DVT) in young women starting the oral contraceptive pill? All these are examples of the clinical features that can be seen in the antiphospholipid syndrome (APS) or Hughes' syndrome.

The background to discovery

In 1983 while studying patients with lupus, we reported a syndrome consisting of thrombosis (both venous and arterial), recurrent miscarriages and a number of other features. We found that the syndrome was characterised by the presence of antibodies against phospholipids. So close is the association between the syndrome and antiphospholipid antibodies that many believe there to be a cause and effect link.

- We realised that the syndrome also existed outside of lupus, and called it the antiphospholipid syndrome (or 'primary' antiphospholipid syndrome, APS for short).
- In 1995, my international colleagues honoured the discovery by calling it Hughes' syndrome.

Is it common?

The answer is yes, possibly one of the commonest of the connective tissue diseases with an impact on all aspects of medicine. This extends from psychiatry to cardiology, from obstetrics to neurology.

Why the term 'sticky blood'?

This feature of the disease refers to the danger of both venous and arterial thrombosis, in particular strokes. In pregnancy, thrombosis of the placenta leads to pregnancy loss.

Who may have Hughes'?

Some possibilities include those with:

- Unexpected strokes (up to 20 per cent of strokes in the under-40s).
- Recurrent miscarriages (especially late fetal death). Up to 20 per cent of women with two or more miscarriages have the syndrome, and it is potentially treatable.
- Recurrent migraine.
- Memory loss: probably from patchy cerebral ischaemia.
- Early and unexpected arterial disease.
- Atypical multiple sclerosis.
- Recurrent DVT's or pulmonary embolism.
- Any unexpected thrombosis.

Confirming the diagnosis

There are two routine diagnostic tests.

- Anticardiolipin antibodies — this is a cheap and simple blood test available in all hospital laboratories.
- Lupus anticoagulant, which is a confusingly named clotting test.

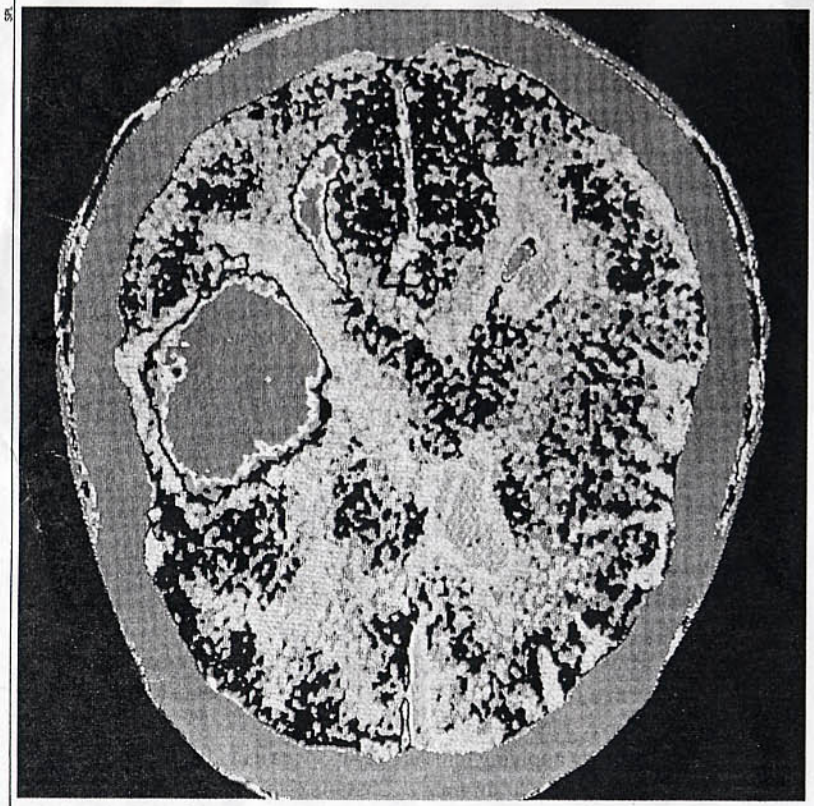
Treating Hughes' syndrome

The 'sticky blood' needs anticoagulants to thin it. The most widely used drug is aspirin in a dose of 75mg to150mg daily.

- Where there has been a major venous thrombosis, or arterial thrombosis, warfarin is vital, and the benefits are sometimes remarkable. For example, in some patients with recurrent headaches or memory loss, there is almost immediate improvement when warfarin is started.
- In recurrent pregnancy loss, the two common regimes are either aspirin 75mg daily or aspirin plus low molecular weight heparin.

How effective is treatment?

It is dramatically effective in some patients. In recurrent pregnancy loss the pregnancy success rate has risen from less than 20 per cent to more than 70 per cent. When treatment is used in the neurological field the impact will be huge. If 20 per cent of strokes in the under 40s are due to this treatable condition, there are huge implications for patients and the NHS.



Cerebral ischaemia (red) symptoms can improve with anticoagulants in Hughes'.

DVT (circled) in young women can be due to Hughes' syndrome.