ment of autonomous parathyroid activity. However, it may also reveal it in those with either prolonged vitamin D deficiency or previously unrecognised primary hyperparathyroidism, a condition now known to be

present in at least 1:1000 of the population. Regular screening for hypercalcaemia in populations receiving such vitamin D repletion regimens is therefore considered to be mandatory.

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Meningismus, fever and macular rash as presenting features of the primary antiphospholipid syndrome?

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Summary

We report here, we believe for the first time, the primary antiphospholipid syndrome, presenting with fever, meningismus and skin rash. Serology was positive for antiphospholipid antibodies but negative for antinuclear factor. Such presentations, once meningitis has been excluded, should be screened for antiphospholipid antibodies. If serology proves to be positive, anticoagulation for life should be considered to avoid thrombotic episodes and death due to pulmonary embolism.

Keywords: antiphospholipid syndrome, meningismus, thrombosis

The antiphospholipid syndrome is a rare but increasingly recognised clinical entity. It was first described by Hughes in 1983¹ as a clinical condition presenting with widespread venous and arterial thrombosis in the presence of antiphospholipid antibodies. Features of this syndrome are widespread and varied, and include stroke, chorea, migraine, transient cerebral ischaemic attack,² livedo-reticularis,³ pulmonary hypertension,⁴ recurrent abortion,⁵ epilepsy, transverse myelopathy, valvular heart disease and ocular ischaemia.⁶ This list will probably be added to, as serological testing for antiphospholipid antibodies is now more general.

We describe the case of a young woman who presented with meningismus, fever and

macular rash and who was found to have positive serology for antiphospholipid antibodies.

Case report

A 21-year-old woman was admitted as an emergency with a 24-hour history of severe generalised headache and photophobia, neck-stiffness and rigors. She had a history of three miscarriages and a previous admission two years previously with headache and meningismus, with a normal computed tomography scan of brain.

Examination revealed an ill-looking woman with a pyrexia of 39.2°C, neck-stiffness, Kernig's sign and a generalised macular erythematous rash. There were no other neurological signs. Initial investigations revealed a polymorphonuclear leukocytosis of $12.1 \times 10^9/l$ (82% polys). C-Reactive protein (CRP) was >150 mg/l (normal range 1–15). Lumbar puncture and other haematological parameters were normal.

Initially a working diagnosis of possible meningococcal meningitis was made and the patient was commenced on high-dose intravenous benzylpenicillin. The following day she was much better and afebrile, but she still had the headache and macular rash. In view of the normal lumbar puncture and clinical improvement, the intravenous antibiotics were stopped. She continued to improve and on the day of discharge was well except for a low-grade pyrexia. Haematological parameters such as white cell count had returned to

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Accepted 6 May 1997

Learning points

- antiphospholipid syndrome can present in a wide variety of ways and is becoming increasingly recognised
- it appears that any age-group can be affected
- antiphospholipid syndrome can occur as a separate entity without features or serological evidence of systemic lupus erythematosus. This is referred to as the primary antiphospholipid syndrome
- treatment may involve long-term anticoagulation to avoid possible fatal thrombosis
- screening of patients with headaches and meningismus may play a vital role in the future for identifying individuals with this syndrome

normal and the CRP was mildly raised (25 mg/l).

Unfortunately, following discharge, she collapsed at home and died. *Post-mortem* examination revealed the cause of death to be a massive pulmonary embolus. Serology carried out showed negative antinuclear factor but was positive for antiphospholipid antibodies. Examination of central nervous system was normal.

Discussion

We believe, due to the constellation of various features, such as three miscarriages, recurrent headaches, and fatal massive pulmonary embolism, that this was an example of the antiphospholipid syndrome. The interest surrounding this case lies in the fact that this

woman presented on two occasions with severe headache and meningismus, with an associated macular rash on the second occasion. Meningismus has been described in patients with systemic lupus erythematosus and positive serology for antiphospholipid syndrome.⁷ However, our patient was positive for antiphospholipid antibodies but had a negative antinuclear factor.

Meningismus is usually associated with inflammation of the meninges and hence there is usually a corresponding change in the cellular constituent in the cerebrospinal fluid. In this case the cerebrospinal fluid examination was normal. We are unable to explain the exact mechanism of the meningismus in this case, but presume that meningeal microvascular thrombosis played a key role. A review by Hughson *et al* concluded that 'the cerebrovascular changes of the antiphospholipid syndrome are derived from a chronic thrombotic microangiopathy'.⁸

This is the first case reported in the literature describing an association between headache, meningismus, macular rash and a positive antiphospholipid antibody. In view of the tragic and fatal event in this case we wish to highlight the possibility that meningismus and the presence of macular rash may be a further manifestation of the antiphospholipid syndrome. We suggest that patients presenting with a meningitic-like illness with macular rash and normal lumbar puncture, should be screened for the presence of antiphospholipid antibodies. Such patients may need serious consideration for appropriate anticoagulation therapy to prevent possible fatal complications.

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