

CASE REPORT

Homonymous hemianopia in the primary antiphospholipid syndrome

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SUMMARY

A woman aged 26 years was referred by her GP to the eye casualty department with sudden-onset left homonymous hemianopia and right-sided headache. Full ophthalmic examination was normal with the exception of a left homonymous hemianopia confirmed with automated perimetry. Urgent CT imaging revealed a non-haemorrhagic cerebral infarct in the right parieto-occipital region. Subsequent blood tests confirmed a diagnosis of antiphospholipid syndrome with positivity in IgG anticardiolipin antibody, IgG anti- β 2-GP1 antibody and the Lupus anticoagulant screen. MRI revealed extensive congenital abnormality at the Circle of Willis, affecting the right half of circulation. The unique dual pathologies may explain her predisposition to right-sided cerebral infarctions. This case highlights a rare but potentially fatal cause of visual disturbance in a young patient, and the importance of the multidisciplinary team approach in diagnosis and management.

CASE PRESENTATION

A woman aged 26 years, a teaching assistant, was referred to the eye casualty by her GP with '24 hours of left homonymous hemianopia'. She reported of losing the left side of her vision for the past 1 day and a right-sided headache, consisting of fluctuations of sharp pain on a background of pain like a 'tight band' around her head.

The patient reported a similar week-long episode of left-sided visual field loss without headache 4 years ago, which spontaneously resolved and was never formally investigated. At presentation, she had been taking the combined oral contraceptive pill (COCP) and gabapentin.

INVESTIGATIONS

On examination, the patient had a blood pressure of 189/112 mm Hg, heart rate 77 bpm and no focal weakness was found. Eye examination revealed discomfort on movement for the left eye. Eye movements were otherwise full with no symptom of diplopia. Visual acuity and pupillary reflexes were normal with no relative afferent pupillary defect. Automated perimetry confirmed the left-sided homonymous hemianopia (figure 1).

Urgent CT head scan performed the same day revealed a right-sided ischaemic parieto-occipital infarct.

Lumbar puncture was performed and the results were unremarkable. Magnetic resonance angiogram

BACKGROUND

Our case highlights the importance of prompt assessment and investigations in patients presenting with 'red-flag' symptoms such as sudden-onset visual field loss, including urgent neuroimaging. It also stresses that antiphospholipid syndrome (APS) is relatively common (20–25%) in young patients with cerebrovascular disease.

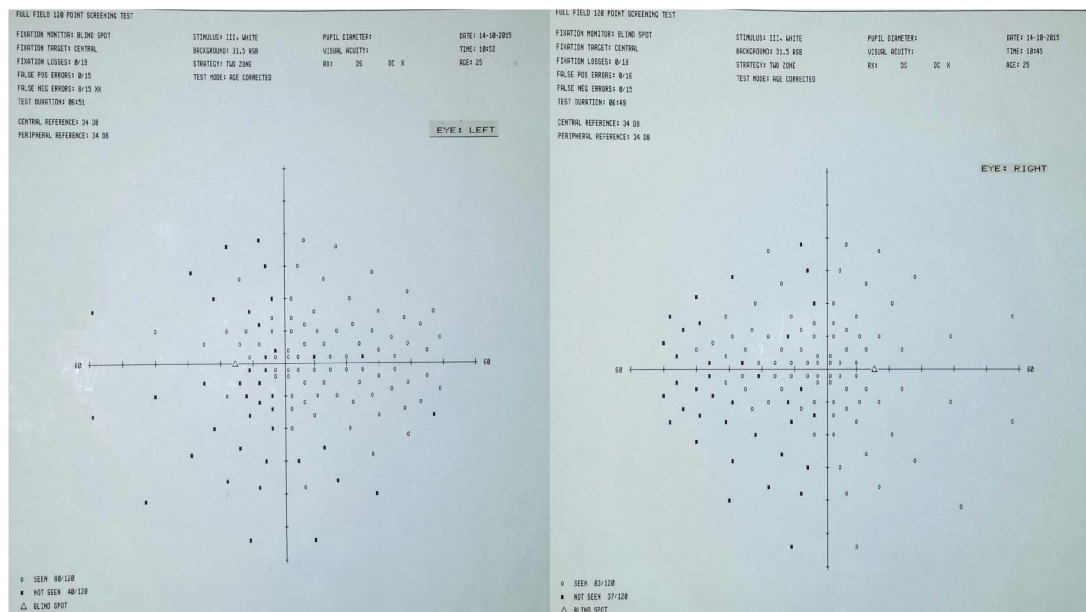


Figure 1 Automated perimetry confirming dense left homonymous hemianopia.



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ruled out venous sinus thrombosis and revealed an acute occipito-cortical infarct with extensive abnormality of the Circle of Willis (figures 2 and 3), including:

- ▶ partial right fetal posterior cerebral artery,
- ▶ absent right middle and anterior cerebral arteries,
- ▶ right posterior cerebral artery arising from the carotid artery.

Blood tests demonstrated positivity in IgG anticardiolipin antibody, IgG anti- β 2-GP1 antibody and the Lupus anticoagulant screen.

DIFFERENTIAL DIAGNOSIS

Other causes of ischaemic or haemorrhagic stroke; arteriovenous malformation.

TREATMENT

While as an inpatient, she was given heparin and subsequently warfarin to reduce the risk of subsequent vascular thrombosis. On discharge, she was advised to continue on lifelong warfarin therapy and to switch from COCP to progestogen-only pill.

OUTCOME AND FOLLOW-UP

The patient was initially admitted to the stroke ward after CT head results. She reported gradual improvement of the visual field defect since admission, which completely resolved after 2 weeks. On discharge, she was continued on warfarin therapy and received follow-up from haematology department as an outpatient.

DISCUSSION

Visual disturbance is a common presentation to eye casualty, and may occasionally represent an underlying systemic disorder.^{1–3} Our case highlights the importance of prompt

assessment and investigations in patients presenting with ‘red-flag’ symptoms such as sudden-onset visual loss.

APS is an autoimmune disease characterised by vascular thrombosis, pregnancy morbidity and presence of antiphospholipid antibodies (box 1).⁴ It typically affects young women and may be primary, or secondary to various rheumatological conditions, including systemic lupus erythematosus and rheumatoid arthritis. Clinical features are systemic and related to hypercoagulability. Stroke, venous sinus thrombosis and retinal vascular occlusion are features of APS that may cause sudden-onset loss of vision.⁵ Of these, stroke is the most common clinical manifestation, with an estimated frequency of 14% among APS patients.⁶ Indeed, stroke in a young patient should raise suspicion of APS—studies estimate that 20–25% of patients under the age of 50 presenting with cerebrovascular disease suffer from APS.^{7–8} Diagnostic criteria involve detection of antiphospholipid antibodies (lupus anticoagulant, anticardiolipin or anti- β 2-glycoprotein 1 antibodies) and the presence of clinical features, namely vascular thrombosis or pregnancy morbidity (box 1).

Treatment is preventive and involves lifestyle advice (regular exercise and healthy diet, avoid smoking and alcohol, reducing cardiovascular risks) and lifelong anticoagulation to a target INR of 2.0–3.0. In addition to thrombosis and pregnancy morbidity, APS is associated with many serious complications, including coronary artery disease, cardiac valvular disease and haemolytic anaemia. Rarely, catastrophic APS results in multiple organ infarctions over a short period of time, necessitating intense anticoagulation and immunosuppression. It is important that APS patients are followed up regularly to ensure adequate anticoagulation.

Box 1 Diagnostic criteria for antiphospholipid syndrome (APS); at least one clinical and one laboratory criterion must be satisfied for a diagnosis of APS⁴

Clinical criteria—at least one of:

- ▶ One or more arterial or venous thrombosis
- ▶ One or more unexplained miscarriages of morphologically normal fetus >10th week of gestation
- ▶ Three or more miscarriages prior to 10 weeks’ gestation
- ▶ One or more late miscarriages or preterm births before 34 weeks’ gestation due to placental insufficiency or pre-eclampsia

Laboratory criteria—at least one of the following, detected on two separate occasions, 12 weeks apart:

- ▶ Anticardiolipin IgM or IgG antibodies (moderate to high titre)
- ▶ Anti- β 2-glycoprotein 1 IgM or IgG antibodies (high titre)
- ▶ Lupus anticoagulant

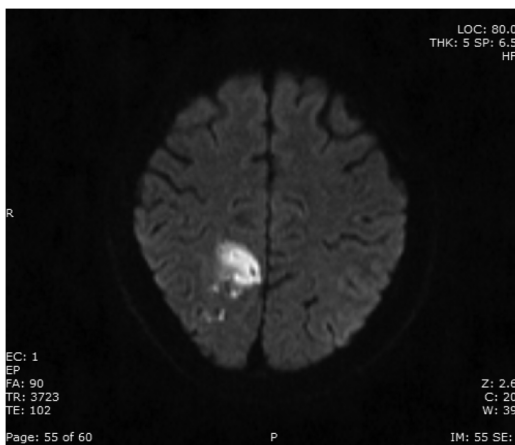


Figure 2 MRI B1000 showing right acute occipito-cortical infarct.

Figure 3 Three-dimensional reconstruction of Circle of Willis displaying abnormal vasculature as described.



Learning points

- ▶ Presentation of homonymous field defects should prompt urgent neuroimaging.
- ▶ Thorough assessment and investigations are necessary for patients with sudden-onset visual loss.
- ▶ Antiphospholipid syndrome (APS) is relatively common in young patients with cerebrovascular disease.
- ▶ APS is a prothrombotic systemic condition, requiring long-term prophylaxis and follow-up.

Contributors DK-HH and JPM conceived of the study. DK-HH and MG collected data for the study. DK-HH and RR performed data interpretation and drafted the article. All authors contributed to the refinement and approval of the final manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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