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MR Findings in Primary Antiphospholipid Syndrome

José R. Pulpeiro,¹ José A. Cortés,² Jesús Macarrón,² Roberto Pérez,³ and Mercedes Bal³

Antiphospholipid antibodies (APLA) are immunoglobulins of the IgG and IgM groups with activity against negatively charged phospholipids. Common manifestations of APLA include the presence of lupus anticoagulant and anticardiolipin antibodies, and false-positive Venereal Disease Research Laboratory (VDRL) and rapid plasma reagin (RPR) tests [1, 2]. APLA has been associated with an increased frequency of venous and arterial thromboses, recurrent fetal loss, thrombocytopenia, cardiac valve lesions, livedo reticularis and other skin lesions, and a variety of neurologic syndromes [1–9]. The disease has been reported most frequently in patients with systemic lupus erythematosus (SLE) and lupuslike disease, but also in other clinical entities, including infections, neoplasms, hematologic disorders, AIDS, primary immunodeficiency, collagenosis, and following treatment with certain drugs (especially in those associated with drug-induced SLE) [1, 2].

Recently, a new subgroup of patients who have idiopathic APLA has been described. The diagnostic criteria of this primary antiphospholipid syndrome (PAPS) include a history of venous and/or arterial thrombotic occlusions, recurrent fetal loss or thrombocytopenia, and the presence of lupus anticoagulant, anticardiolipin antibodies, or both, in the absence of SLE or any other cause of APLA [1, 2]. To make the diagnosis, at least one clinical feature and one positive laboratory test should be present on two different occasions separated by a 2-month interval [2]. We report the MR findings in a case of PAPS clinically resembling multiple sclerosis.

Case Report

A 59-year-old woman was admitted to the hospital because of monocular blindness that developed during the previous 5 days. Her history revealed that she had had an episode of left leg weakness 6 years earlier that was associated with extensor plantar responses, brisk deep reflexes, left palpebral ptosis, diminished sensitivity in the right hemithorax and upper limb, and a left cerebellar syndrome. She recovered completely but suffered a new episode of right arm and leg weakness and a right cerebellar syndrome 1 month later.

At this time, laboratory tests were unremarkable except for the presence of mild thrombocytopenia and mild protein elevation in the

CSF. Pre- and postcontrast CT scans of the brain were normal. Multiple sclerosis was clinically suspected.

The patient remained asymptomatic except for mild gait ataxia. On admission, physical examination showed total visual loss in the right eye and slight pallor of the optic disk on fundoscopy. There was also increased tone and hyperreflexia in all four limbs with normal power and abnormal plantar responses. Pain, vibratory, and touch sensations were normal. There was a broad-based unsteady gait. Romberg test was negative, and there was no dysmetria. The patient had no history of skin lesions (including lupus rashes and livedo reticularis), uveitis, oral or genital ulcers, or arthritis; nor were these lesions found on physical examination. Laboratory results showed prolonged partial thromboplastin time, noncorrectable by 1:1 mixture with normal plasma, and prolonged dilute Russell viper venom time, consistent with the presence of lupus anticoagulant. High levels of IgG anticardiolipin antibodies (310 GPI μ /ml) were also present at two different times. Tests for VDRL, RPR, and antinuclear antibodies were negative. There was mild thrombocytopenia. CSF examination, including immunoelectrophoresis, was normal. Visual evoked potentials were abnormal on the right and normal on the left. Brainstem and spinal potentials were normal.

Plain CT revealed calcification in the right caudate nucleus that was considered of no clinical significance. Double-dose delayed post-contrast CT failed to demonstrate white matter demyelination. MR showed multiple subcortical foci of high signal intensity on T2-weighted images (Fig. 1).

On the basis of the patient's history and laboratory data, we made the diagnosis of PAPS and ischemic optic neuritis, and instituted treatment with anticoagulants, which partially improved visual acuity.

Discussion

A close relationship between APLA and neurologic disease has been documented [5–7]. Cerebrovascular disorders (including stroke, transient ischemic attacks, multiinfarct dementia, and acute ischemic encephalopathy) are reported most frequently, with a rate of occurrence of 25–31% [6, 7]. Transient ischemic attacks and stroke account for most cases of arterial thromboses associated with APLA and tend to occur in patients younger than 45 years [2]. Symptoms of ocular ischemia, such as amaurosis fugax, and retinal artery or vein thrombosis have also been described [6, 9]. Reported cases of APLA-associated noncerebrovascular neurologic diseases include SLE, Jamaican and Degos disease myelop-

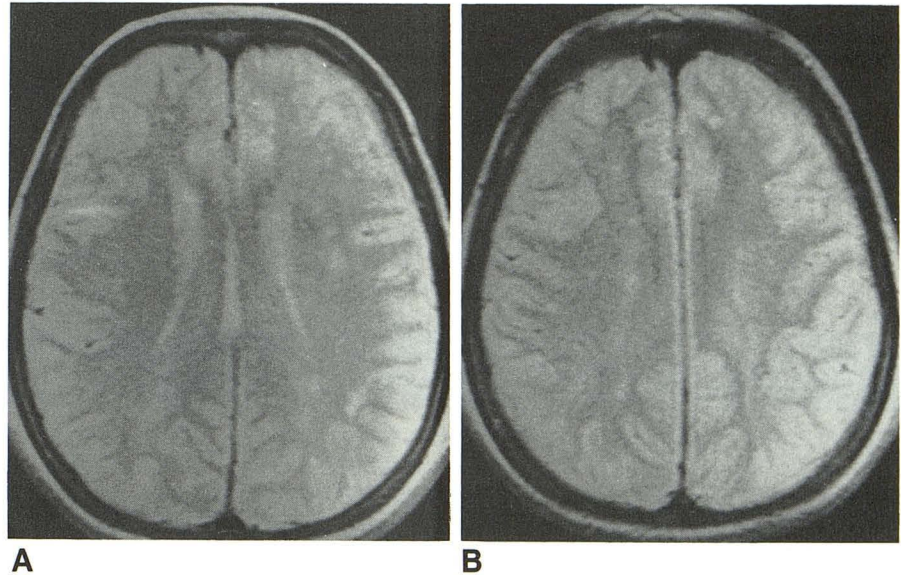
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¹ Department of Radiology, Hospital Xeral, c/ Premio Nobel Dr. Ochoa, s/n, 27004 Lugo, Spain. Address reprint requests to J. R. Pulpeiro.

² Department of Neurology, Hospital Xeral, Lugo, Spain.

³ Department of Internal Medicine, Hospital Xeral, Lugo, Spain.

Fig. 1.—A and B, Spin-echo MR images (2208/50/2) show bilateral periventricular and subcortical small high-signal foci in white matter.



athy, Guillain-Barré syndrome, migraine, chorea, seizures, and a syndrome resembling multiple sclerosis (pseudomultiple sclerosis) [5, 8].

The pathogenesis of these APLA-associated neurologic disorders is poorly understood. In most cases the neurologic symptoms reflect ischemic arteriopathy in medium-size vessels. Stenosis and occlusions in medium-size arteries have been described angiographically [5], and platelet-fibrin thrombi filling small vessels without signs of vasculitis have been histologically demonstrated in some cases [6].

The mechanism of this APLA-related thrombosis is unknown. Direct neural damage mediated by cross-reactivity of APLA with CNS phospholipids may also be implicated in the pathogenesis of neurologic disease [5, 8], and high levels of APLA, especially of the IgG class, appear to be associated with more severe neurologic disease [2, 5].

The CT findings in patients with APLA (and those with PAPS) and neurologic impairment include diffuse cerebral atrophy and solitary or multiple small hypodense lesions in cortical, periventricular, and subcortical regions. CT studies may also be normal [7]. Subcortical bright foci on MR images have also been described [9]. These lesions probably represent small areas of infarction or demyelination, but to our knowledge, no radiologic-pathologic correlation is available to date. Similar lesions have been reported in many other CNS diseases, including multiple sclerosis, SLE, Behçet disease, Sjögren syndrome, Sneddon syndrome, migraine, degenerative microangiopathy, and multifocal infarcts [10, 11]. APLA has also been found in some of these disorders [1, 2]. The presence of APLA may thus provide a common link in the pathogenesis of some acquired white matter disorders and may define new subgroups among them, such as PAPS. Confirmation of the presence of APLA may also clarify some cases of clinically suspected multiple sclerosis or idiopathic

strokes in young patients, and probably should be made in such cases. Because lupus anticoagulant has been reported to be present in only 50–90% of patients who have anticardiolipin antibodies and lupus anticoagulant can be present in the absence of anticardiolipin antibodies [5, 6, 9], both assays should be performed to reliably detect APLA.

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