and medium-sized arteriopathy. On the basis of these findings, they suggested that the pathogenesis of cerebral ischemia in Sneddon’s syndrome differs depending on whether aPL antibodies are present or absent, namely, large-vessel arteriopathy in aPL antibody-positive patients and medium-vessel arteriopathy in aPL antibody-negative patients. In his response, Levine stated that the difference in the size of the vessels affected in Sneddon’s syndrome is not so clear cut. We report the case of a patient with typical Sneddon’s syndrome, high levels of aPL antibodies, medium-sized arteriopathy, and lack of involvement of large cerebral vessels.

A 44-year-old white man was admitted to the hospital in April 1989 because of left basal pneumonia. His previous history was unremarkable. He was treated with erythromycin with good recovery. Antibody titer for Legionella pneumophila became elevated from 1/1,024 to 1/4,096. One month later livedo reticularis appeared on all four limbs and his trunk. Two months after that, he developed a mild left hemiparesis with sensory impairment, but no other neurological sign. Livedo reticularis was still present. His neurological condition recovered in 4 days. Cranial computed tomography, electrocardiogram, four-vessel angiogram, and routine laboratory tests were normal. The following antibody titers were negative: antiglutaraldehyde, anti-DNA, anti-ENA, anti-SSA, and anti-SSB. A skin biopsy was normal. Rapid plasma reagin test was positive. High levels of antiphospholipid antibodies were found (30 GPL and 10 MPL). An echocardiogram showed a slightly incompetent and fibrosed aortic valve. Brain magnetic resonance imaging, on the T2-weighted sequences, showed small, hyperintensive lesions on the right frontoparietal white matter, and digital artery biopsy showed focal areas of intimal hyperplasia and a redundant elastic internal layer. Treatment with aspirin was started (500 mg, once a day). The evolution of different aPL antibody levels is shown in Table 1. Although livedo reticularis persisted, no further ischemic cerebrovascular events took place during a 20-month follow-up period.

A noninflammatory arteriopathy, affecting cerebral and cutaneous vessels, has been reported in Sneddon’s syndrome. Its pathogenesis is unknown, but there are some reports of Sneddon’s syndrome associated with aPL antibodies, suggesting that these antibodies may play a role. Nowadays, it seems clear that the association of livedo reticularis with cerebrovascular lesions could be the result of a wide spectrum of causes, one of which is the “primary” antiphospholipid syndrome.

We feel that the differences in immunological and histological findings from case to case could reflect clinical heterogeneity, divergences in laboratory methods employed in aPL investigations, or both. These dissimilarities stress the necessity of homogeneity in the study of these patients.
The following is in response:

To the Editor:

In our report1 of a patient with Sneddon's syndrome and negative antiphospholipid antibodies, we said that there were angiographic differences between Sneddon's syndrome and cerebrovascular lesions associated with antiphospholipid antibodies. While cerebral angiography in patients with cerebrovascular disease and antiphospholipid antibody syndrome has either been normal or revealed large-vessel occlusions,2 middle-sized arteries involved in Sneddon's syndrome can be drawn from immunologically, and histologically heterogeneous, partly because of the nonspecific nature of livedo reticularis and cerebral infarction. Antiphospholipid antibodies represent only one common association between the two clinical phenomena.3

I am not sure that the pathogenesis of cerebral ischemia in Sneddon's syndrome was different whether antiphospholipid antibodies were present or not.

Moral et al report a patient with Sneddon's syndrome and positive antiphospholipid antibodies with biopsy evidence of medium-sized arteriopathy. Sneddon's syndrome is characterized by the presence of a noninflammatory medium-sized arteriopathy. Digital artery biopsy has shown intimal hyperplasia in nearly all patients in whom this procedure was performed.1,2 The case reported by Moral et al supports the hypothesis that this occlusive arteriopathy is the main factor involved in the pathogenesis of cerebrovascular lesions in Sneddon's syndrome.

The significance of antiphospholipid antibodies in Sneddon's syndrome is still unclear although Kalashnikova et al8 found that their presence is associated with a poorer outcome. Finally, we agree with Moral et al on the need for standardized laboratory methods in the study of antiphospholipid antibodies in patients with Sneddon's syndrome.

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Stroke. 1991;22:1327-1328
doi: 10.1161/01.STR.22.10.1327

Stroke is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0039-2499. Online ISSN: 1524-4628

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