Sneddon's Syndrome

- A Case Report -

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Sneddon's syndrome consists of widespread livedo reticularis and ischemic cerebral manifestations. We report a case of a 70-year-old Korean woman with extensive livedo reticularis, hypertension, positive laboratory results for antinuclear antibodies and the lupus anticoagulant test, and idiopathic Parkinson's disease. (Ann Dermatol 11(1) 62~64, 1999).

Key Words : Sneddon's syndrome, Livedo reticularis, Ischemic cerebral manifestation

Sneddon's syndrome is an infrequent neurocutaneous disorder of unknown origin first described by Ehrmann in 1907 and recognized as a separate entity by a British dermatologist, Sneddon in 1965[1]. Sneddon[2] described 6 patients with cerebrovascular incidents which have been of limited and benign nature, often leaving little residual disability , associated with a benign type of hypertension and livedo reticularis. The neurological events first described were motor deficits, aphagia and visual field defects[3], and the most common neurological manifestations consist of cerebrovascular accidents, transient ischemic attacks, convulsions and/or progressive dementia[4]. Also, pateints with Sneddon's syndrome have been reported to have antiphospholipid antibodies[5]. Recently, the systemic nature of this syndrome has been described the typical characteristic vascular lesions are located in the renal area[4].

We present a patient with extensive livedo reticularis, hypertension, ischemic cerebral manifestations and Parkinson's disease.

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CASE REPORT

A 70-year-old Korean woman with idiopathic Parkinson's disease was referred from the neurology department for the problem of extensive livedo reticularis on both the forearms and thighs. She was diagnosed with essential hypertension 2 years previously, and developed bradykinesia and gait disturbances one year previously. The neurologic examination demonstrated multiple signs of Parkinson's disease: bradykinesia, rigidity, gait difficulty and a mask face. Tremors were not noticed. Results of motor and sensory examinations were normal for her age. There was a family history of hypertension. However, she denied a history of spontaneous abortion and of skin disease in her family members.

Laboratory examinations revealed the following abnormalities: the antinuclear antibody (ANA) test result was positive at 1:160 in a homogenous pattern and the lupus anticoagulant (LAC) test was positive. Other laboratory test results including a complete blood cell count, erythrocyte sedimentation rate, liver and lipid profiles, thyroid function test, coagulation battery, protein C, protein S, fasting blood glucose, anticardiolipin antibodies (IgG/IgM), extractable nuclear antigens (ENA) profiles, anticytoplasmic neutrophilic antibodies (ANCA), cryoglobulin, cold agglutinin and urinalysis were negative or within normal limits. Brain magnetic resonance imaging (MRI) re-

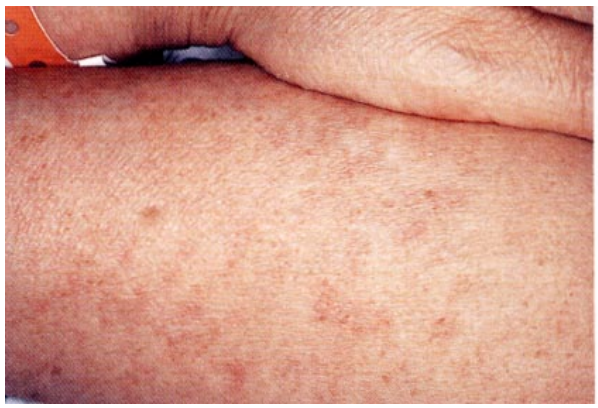

Fig. 1. Extensive, confluent erythematous, reticulated patches on the forearms.

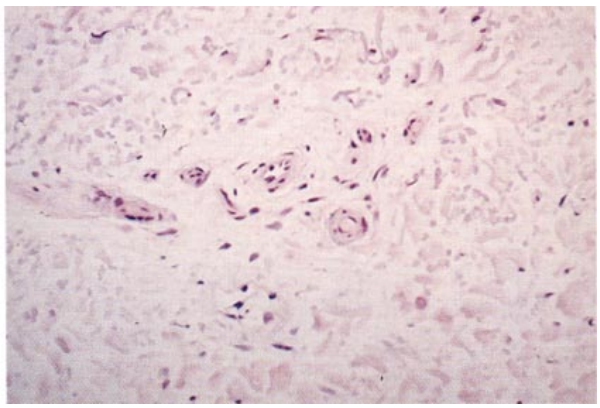

Fig. 2. Thickened small blood vessels in the upper dermis (hematoxylin and eosin, × 100).

vealed diffuse multiple lacunae in the cerebral lobes. MR angiography showed normal result. Brain IPT (N-(3-iodiopropen-2-yl)-2-carbomethoxy -3(-(4-chlorophenyl)-tropane)) single photon emission computed tomography (SPECT) revealed a severe decreased uptake in both basal ganglia. Skin examinations showed extensive, confluent, reticulated, erythematous patches on both the forearms and thighs (Fig. 1). The skin lesions had developed three years previously with a progressive nature. A biopsy specimen from the forearm revealed that the vessles in the upper dermis were thickened and slightly obliterated and the dermis showed actinic degeneration (Fig. 2). No inflammatory cells were identified around or within the vessel.

DISCUSSION

Livedo reticularis occurs in connective tissue diseases such as systemic lupus erythematosus (SLE), thrombotic disorders such as thrombotic thrombocytopenic purpura, idiopathic thrombocytopenia, and atheromatous vascular disease, infectious illness including syphilis and tuberculosis, other immunological conditions including polyarteritis nodosa, cryoglobulinemia, and in cholesterol embolization, and as a side effect of drugs such as amantadine[5-7]. About half of the cases are idiopathic[8].

Although our patient did not fulfill the American Rheumatism Association's criteria for SLE and the criteria for antiphospholipid antibody syndrome, positive laboratory results for ANA (1:160, homogenous pattern) and the LAC test were interesting. The associations between Sneddon's syndrome, antiphospholipid antibody syndrome and incipient SLE were reported[8,9]. The mechanisms by which antiphospholipid antibodies theoretically contribute to ischemic brain and skin disorders include inhibition of phospholipid-dependent endogenous anticoagulants, direct vascular endothelial damage, and antibody-mediated platelet activation, all of which can promote thrombosis[10]. Of the patients with Sneddon's syndrome who had been screened for antiphospholipid antibodies, the rates of positivity ranged from zero to 86%.[11,12] Zelger et al.[1] described that the presence or absence of antiphospholipid antibody was not an exclusion or an inclusion criterion for Sneddon's syndrome and hypertension is the only risk factor significantly associated with a more severe course of the disease. The analysis of skin biopsies from 21 patients of Sneddon's syndrome led Zelger et al.[1] to suggest a history of cutaneous vascular events based on the endothelial target as a primary event and to speculate on possible similar vascular lesions in the central nervous system. So, recently Tourbach et al.[3] reported a retrospective analysis of the neurological events. Biological findings including antiphospholipid antibodies, and MRI findings were present in 26 patients with Sneddon's syndrome. They concluded that the severity of the diseases seems to be correlated with brain MRI aspects, but not to the presence of antiphospholipid antibodies, the presence of hypertension or other vascular risk factors. Any-

way, the positive results of ANA and LAC in our patient might support the previous reports about the associations between this syndrome and incipient SLE and antiphospholipid antibody syndrome.

In our patient, brain MRI revealed multiple lacunae, which were attributed to thrombosed cerebral vessels in Sneddon's syndrome. MR angiography was normal and IPT brain SPECT showed severely decreased uptakes in both basal ganglia, which led to the diagnosis of the idiopathic Parkinson's disease based on the neurological findings. Although the association between the idiopathic Parkinson's disease and Sneddon's syndrome may be conjectured, we concluded those two diseases coexisted because the idiopathic Parkinson's disease of which pathophysiology is still a mystery, did not develop from vascular disorders. In Korea, only 2 cases were described in Neurology[13,14]. In both cases, Sneddon's syndrome was related to cerebral infarction.

We report a case of Sneddon's syndrome with characteristic clinical and histopathological findings of livedo reticularis, hypertension, and multiple lacunae in brain MRI, positive laboratory results for ANA and LAC, and idiopathic Parkinson's disease. The awareness of this rare syndrome should be made in considering the neurological impairments in patients with livedo reticularis.

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