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Case Report

Antiphospholipid Antibody Positive Sneddon Syndrome: a Case Report

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Abstract

Sneddon syndrome may present with neurological findings such as transient ischemic stroke, strokes, seizures and/or headaches. However, a purplish, spider web-like skin finding called livedo reticularis may accompany the skin and precede neurological findings. Sneddon syndrome often affects women. Since it is vasculopathy affecting small and medium vessels, other organ findings may accompany. We present a 44-year-old Sneddon syndrome patient with monoparesis in her left lower extremity, livedo reticularis on her back and legs, and hypertension.

Keywords

antiphospholipid, Sneddon, stroke, warfarin

INTRODUCTION

Sneddon syndrome is a rare occlusive arterial disease characterized by recurrent ischemic stroke and livedo reticularis in the skin. It usually affects young adults and women with an average age of onset between 20 and 42 years.[1-3] Stroke, transient ischemic attacks, seizures, headache, and dizziness may be seen among the neurological findings in Sneddon syndrome. Since small and medium-sized arteries are involved in the central nervous system, clinical findings may not be very severe. Although the etiology of Sneddon syndrome is not generally known, it may sometimes be associated with an autoimmune disease.[4-6] Most cases are sporadic, but some familial cases with autosomal dominant inheritance have been reported.[4-7] It has been emphasized in the literature that Sneddon syndrome should be differentiated according to the positivity or negativity of antiphospholipid antibodies. Its incidence has been reported to be 4 per 1 million per year in the general population.[1-3,5] Livedo reticularis is a purplish skin change with a spider web-like appearance that can be seen on the trunk and extremities. It can be seen before a stroke. Livedo reticularis can worsen cold and pregnancy. Livedo reticularis may cause diseases such as some collagen tissue diseases (systemic lupus erythematosus, polyarteritis nodosa, rheumatoid arthritis), metabolic disorders (hypercalcemia), infections (tuberculosis, syphilis), hematological disorders (thrombotic thrombocytopenic purpura, antiphospholipid antibody syndrome, disseminated intravascular coagulation), cryoglobulinemia. These may accompany the condition and should be considered in the differential diagnosis.[4-6] Hypertension occurs in a significant proportion of patients with Sneddon syndrome. Central retinal artery occlusion and central retinal vein occlusion can be seen among its ophthalmological complications.[1,2,5,7]

CASE REPORT

A 44-year-old right-handed female patient presented with complaints of numbness and weakness in her left leg that had been present for about a day. She had no disease oth-

er than known hypertension for which she was taking lercadipine 20 mg. On physical examination, her blood pressure was 142/70 mmHg, heart rate was 88/min, temperature was 36.5°C, and oxygen saturation was 97% in room air. Electrocardiography (ECG) showed a normal sinus rhythm. There were purple-colored skin lesions on her back that extended to her hips, which were not raised from the skin, similar to spider webs, and fading with pressure. The neurological examination of the patient revealed left lower extremity strength of -5/5 and hypoesthesia in the left lower extremity. Deep tendon reflexes were normoactive. The patient had occasional double vision for ten years and had a history of abortion in 2012. No pathological finding was detected by the brain computed tomography (CT). Magnetic resonance imaging (MRI) of the patient showed diffusion restriction in favor of acute-subacute infarct, located paramedian at the vertex level in the right frontoparietal, and involving cortical-subcortical areas ***(Fig. 1)****.*

In the dermatology consultation of the patient, the skin finding was evaluated as livedo racemosa and biopsy was taken ***(Fig. 2)****.*

No significant pathology or cardioembolic stroke focus was detected in the cardiac examination. No vessel occlusion was found in the brain-neck CT angiography.

In laboratory tests, complete blood count, platelet function tests, coagulation tests, routine biochemistry tests, complete

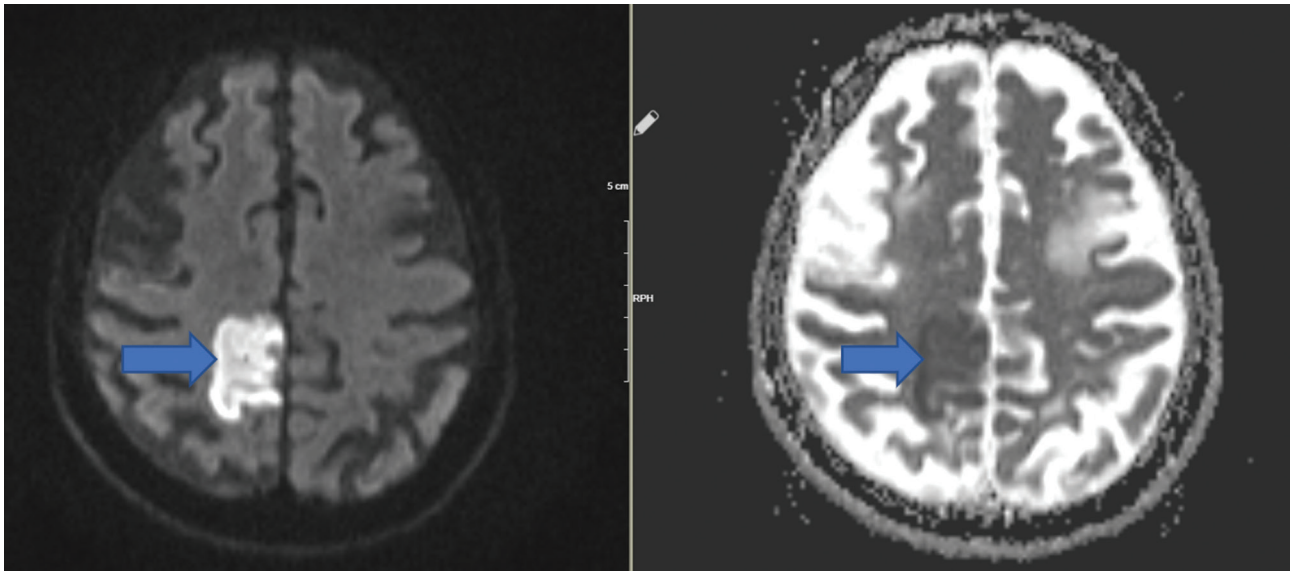

Figure 1. Acute infarct located paramedian at the vertex level in the right frontoparietal vertex in diffusion MRI imaging, involving cortical-subcortical areas, and its counterpart in ADC sequence.

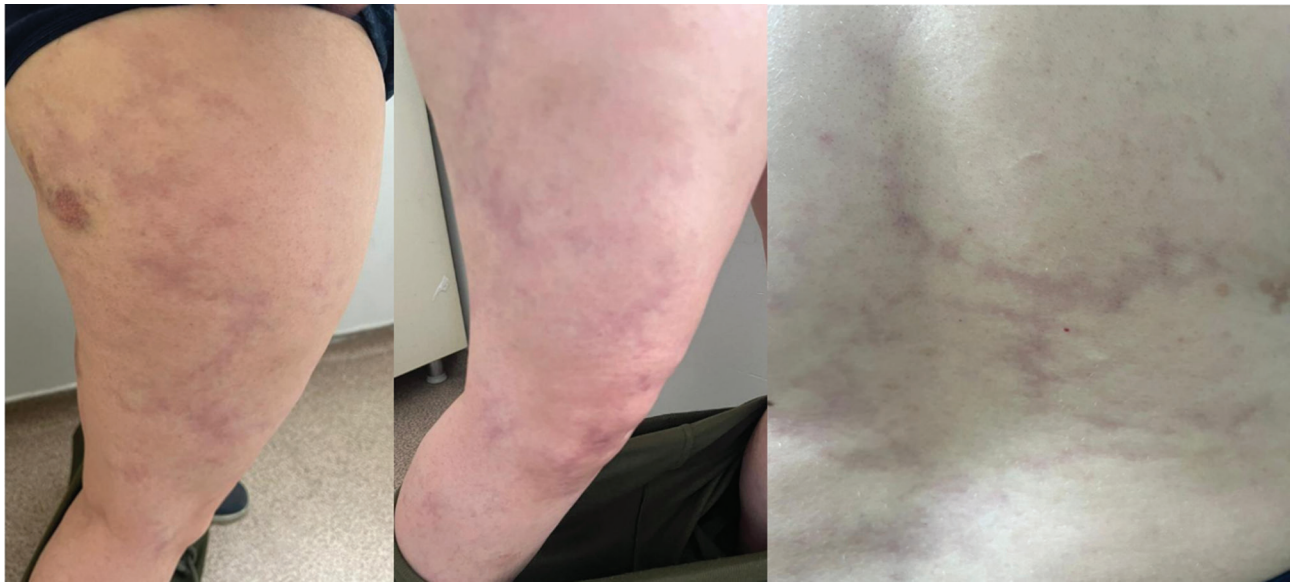

Figure 2. Spiderweb-like, purplish, non-fading livedo reticularis skin lesions on the legs and back of the patient

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urinalysis, protein electrophoresis, antinuclear antibodies, vitamin B12, thyroid function tests, HbA1c, erythrocyte sedimentation rate, autoantibody (anti-SSA, anti-SSB, anti-dsDNA antibodies, anti-Sm antibodies), antithyroid antibodies, syphilis serology (fluorescent treponemal antibody), Schirmer test, homocysteine, rheumatoid factor, anti-streptolysin O (ASO), rheumatoid factor (RF), immunoglobins A, G, and M were normal. ELISA tests (hepatitis A, HIV, hepatitis B, hepatitis C) were negative. However, anti-cardiolipin IgM elevation was detected but anti-β2 glycoprotein 1, anti-prothrombin IgG and IgM, anti-annexin V IgG, and anti-annexin V IgM were normal. The tests for viral meningitis agents (herpes simplex virus 1-2, HSV), varicella zoster virus (VZV), enterovirus, parechovirus, Epstein-Barr virus (EBV), cytomegalovirus (CMV), and adenovirus were negative. The brucella tests were negative. The pathergy test for Behçet's disease was negative. In skin biopsy, proliferating vascular structures under the epithelial tissue and intense mononuclear type inflammatory cell infiltration around these vascular structures were consistent with livedo reticularis. No significant pathology was detected with the renal Doppler ultrasound study for hypertension. No additional pathology was detected in the examinations performed in the patient who had occasional double vision.

Warfarin treatment was administered to the patient due to a history of thrombosis and anticardiolipin Ig M positivity. The patient was discharged from the outpatient clinic to be followed up. However, the patient did not come for examination afterwards.

DISCUSSION

Sneddon syndrome is a rare occlusive arterial disease characterized by recurrent ischemic stroke and livedo reticularis in the skin. Stroke, transient ischemic attacks, seizures, and headaches can be seen in patients as neurological findings. However, patients may have liver, kidney diseases, and hypertension.[1-4] Our patient had hypertension, liver and kidney function tests were normal, and no significant pathology was found by the ultrasonography performed to find the etiology of hypertension. On MRI, an acute-subacute infarct located paramedian at the vertex level in the right frontoparietal and involving cortical-subcortical areas was detected. Thereupon, the etiology of ischemic stroke was investigated in our patient. Sneddon syndrome affects women more frequently between the ages of 20 and 42 and its incidence is estimated to be 4 in 1 million people per year.[1-3,5] Our patient was a 44-year-old woman whose biopsy result of the skin findings was found to be compatible with livedo reticularis. Some collagen tissue diseases, metabolic disorders, infections (tuberculosis, syphilis), hematological disorders that may be associated with livedo reticularis were excluded.[4-6] The relationship between cerebrovascular disease and livedo reticularis was reported in 1959 by Kimming and described by Champion and Rook in 1960. The syndrome, however, was first described by Sneddon in 1965.[1-3,7] Antiphospholipids are associated with a high risk of ischemic stroke, and they were positive in our patient. As a result of the thrombophilia panel searched in our patient, no significant pathology was detected.

Treatment with warfarin is recommended in the presence of antiphospholipid antibodies.[1-3,7] Our patient had antiphospholipid antibodies and warfarin treatment was started.

We wanted to present our 44-year-old patient with ischemic stroke, livedo reticularis and hypertension, anti-cardiolipin IgM positivity and a diagnosis of Sneddon syndrome.

In conclusion, Sneddon syndrome, which is very rare, was aimed to be considered while investigating the etiology in young stroke patients. If the patient meets the diagnostic criteria for Sneddon syndrome, antiphospholipid antibodies and thrombophilia panel tests should also be performed in order to regulate the treatment, and the treatment should be arranged accordingly.

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Competing Interest

The authors declare that they have no known competing financial or personal relationships that could have appeared to influence to work reported in this paper.

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Синдром Sneddon с положительными антифосфолипидными антителами: описание случая

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Резюме

Синдром Sneddon может проявляться неврологическими проявлениями, такими как преходящий ишемический инсульт, инсульты, судороги и/или головные боли. Тем не менее, пурпурное, похожее на паутину образование на коже, называемое *ливедо ретикуларис*, может обнаружиться на коже и предшествовать неврологическим симптомам. Синдром Sneddon часто поражает женщин. Поскольку это васкулопатия, поражающая мелкие и средние сосуды, она может сопровождаться изменениями в других органах. Мы представляем 44-летнюю пациентку с синдромом Sneddon с монопарезом левой нижней конечности, ливедо ретикуларис на спине и ногах и гипертонией.

Ключевые слова

антифосфолипид, Sneddon, инсульт, варфарин