VASCULAR DISEASE, VASCULITIS

A CASE OF SNEDDON'S SYNDROME

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Background: Sneddon's syndrome (SS) is a rare, non-inflammatory thrombotic vasculopathy with an incidence around 4/1.000.000 habitants. It mostly affects women between 20 and 42 years of age. The SS is increasingly recognised as a cause of ischaemic stroke in young adults.

Observation: A 41 years old woman came to our attention for severe disabling headache, transient aphasia and visual defects. The patient was afebrile with blood pressure 120/80 mmHg and medium frequency sinus rhythm, without memory cognitive deficits and autonomous in Activities Daily Living (ADL). She had hypertension in treatment with ACE inhibitors. Physical examination revealed a dusky erythematous-violaceous, irregular, lacelike pattern in the skin of abdomen and breast (appeared four years earlier) as livedo reticularis (subsequently confirmed by skin biopsy). Blood tests showed mild anemia. She underwent head CT scan with contrast that revealed mild atrophy of white matter.

Key message: Sneddon's syndrome is a rare disorder characterized by the occurrence of cerebrovascular disease associated with livedo reticularis. Pathophysiology of SS is unclear although various abnormalities have been reported in isolated cases: activated protein C resistance, platelet aggregability, increased thromboglobulin levels, familial deficiency in antithrombin III and protein S deficiency, modifications of the ratio tissue plasminogen activator/inhibitor, antiphospholipid antibodies. It is speculated that a nonvasculitic small and medium sized vessel arteriopathy cause both skin symptoms and cerebrovascular events. The treatment of SS is controversial but long-term anticoagulation is recommended. It is important to recognize the Sneddon's syndrome, particularly because stroke episodes may be prevented through appropriate treatment.