

## “My skin’s all blotchy!”

Benjamin Barankin, MD

A 48-year-old male presents with blotchy skin, predominantly on the trunk, but, more recently, progressing to include the arms and legs. The blotchy skin is fixed, and not affected by temperature. He has a history of multiple strokes for which a cause has not been found. There is no family history of clotting problems.

### What is your diagnosis?

This gentleman has *livedo reticularis* (fish-net vascular appearance) as part of Sneddon syndrome. Sneddon syndrome is a rare disorder of unknown etiology characterized by widespread *livedo reticularis* and multiple strokes, resulting in dementia.

The incidence is estimated at four cases per million people a year.

Some patients with this condition have antiphospholipid antibodies and anti-β<sub>2</sub>-glycoprotein antibodies, while others have antibodies of systemic lupus erythematosus.




Figure 1. Sneddon syndrome.

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nerves, heart and kidneys is frequent (50% to 70%), yet usually asymptomatic. Hypertension is the only risk factor significantly associated with a more severe course of the disease and, unfortunately, medications are often ineffective at controlling it.

Non-specific prodromal symptoms, such as headache and dizziness, often (80%) precede the livedoid skin pattern. The cutaneous manifestation of *livedo reticularis* or *livedo racemosa* may precede the cerebrovascular episodes, thus alerting the clinician; it appears to be an irreversible skin finding.

Multiple skin biopsies are often needed to reveal inflammatory findings ("endothelitis") of small- to medium-sized arteries followed by subendothelial proliferation and fibrosis and arteriolar occlusion.

The success of warfarin in the treatment of antiphospholipid syndromes and the failure of immunosuppression and acetylsalicylic acid in the treatment of Sneddon's syndrome suggest that warfarin anticoagulation may currently be the most appropriate intervention.

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