ONLINE EXCLUSIVE

White ankle scars

We discovered that these white lesions were remnants of a deeper issue that linked back to significant events in the patient's medical past.

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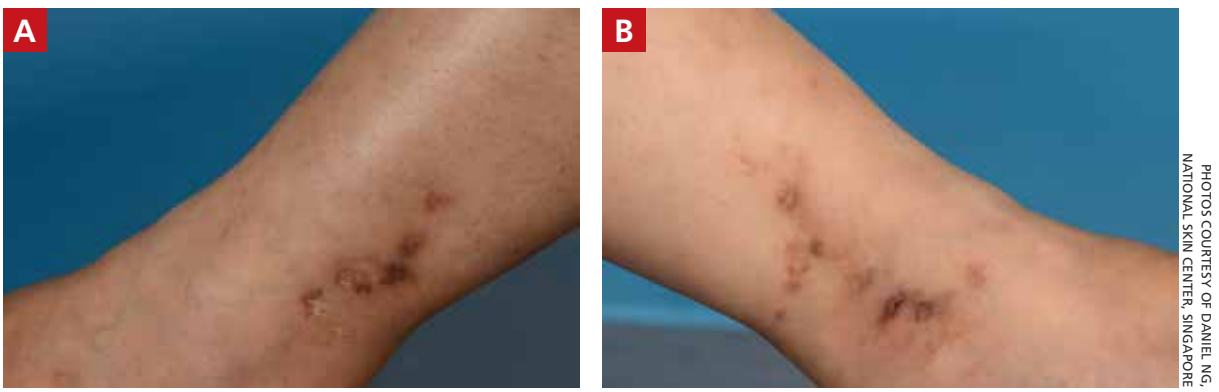
A 42-YEAR-OLD WOMAN presented to our dermatology center with white scars on both of her ankles. She first noticed the lesions 2 years prior; they were initially erythematous and painful, even when she was at rest. Her past medical history included 3 spontaneous term miscarriages. She denied any prolonged standing or trauma.

On examination, atrophic porcelain-white stellate scars were visible with surrounding hyperpigmentation on the medial aspect of both ankles (**FIGURE 1A & 1B**). There were no tender erythematous nodules, livedo reticularis, varicosities, or pedal edema present. The dorsalis pedis pulse was well felt and capillary refill time was less than 2 seconds; sensation was intact.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE 1
Atrophic white stellate scars with surrounding hyperpigmentation on both ankles



PHOTOS COURTESY OF DANIEL NG, NATIONAL SKIN CENTER, SINGAPORE

Atrophie blanche is an important clue to acquired thrombophilia.

Diagnosis: Atrophie blanche

Atrophie blanche is a morphologic feature described as porcelain-white stellate scars with surrounding telangiectasia and hyperpigmentation. The lesions are typically found over the peri-malleolar region and are sequelae of healed erythematous and painful ulcers. The lesions arise from upper dermal, small vessel, thrombotic vasculopathy leading to ischemic rest pain; if left untreated, atrophic white scars eventually develop.

A sign of venous insufficiency or thrombotic vasculopathy

Atrophie blanche may develop following healing of an ulcer due to venous insufficiency or small vessel thrombotic vasculopathy.[1] The incidence of thrombotic vasculopathy is 1:100,000 with a female predominance, and up to 50% of cases are associated with procoagulant conditions.[2] Thrombotic vasculopathy can be due to an inherited or acquired thrombophilia.[1]

Causes of hereditary thrombophilia include Factor V Leiden/prothrombin mutations, anti-thrombin III/protein C/protein S deficiencies, dysfibrinogenemia, and hyperhomocysteinemia.

Acquired thrombophilia arises from underlying prothrombotic states associated with the Virchow triad: hypercoagulability, blood flow stasis, and endothelial injury. The use of oral contraceptives or hormone replacement therapy, presence of malignancy, and antiphospholipid syndrome (APS) are causes of acquired thrombophilia.[2]

Obtaining a careful history is crucial

Thorough history-taking and physical examination are required to determine the underlying cause of atrophie blanche.

Chronic venous insufficiency is more likely in patients with a history of prolonged standing, obesity, or previous injury/surgery to leg veins. Physical examination would reveal hyperpigmentation, telangiectasia, varicose veins, pedal edema, and venous ulcers.[3]

Inherited thrombophilia may be at work in patients with a family history of arterial and venous thrombosis (eg, stroke, acute coronary syndrome, or deep vein thromboses).

Acquired thrombophilia should be suspected if there is a history of recurrent miscarriages or malignancy.[4] Given our patient's history of miscarriages, we ordered further lab work and found that she had elevated anticardiolipin levels (> 40 U/mL) fulfilling the revised Sapporo criteria[5] for APS.

Thrombophilia or chronic venous insufficiency? In a patient with a history suggestive of thrombophilia, further work-up should be done before attributing atrophie blanche to healed venous ulcers from chronic venous insufficiency. A skin lesion biopsy could reveal classic changes of thrombotic vasculopathy subjacent to the ulcer, including intraluminal thrombosis, endothelial proliferation, and subintimal hyaline degeneration, as opposed to dermal changes consistent with venous stasis, such as increased siderophages, hemosiderin deposition, erythrocyte extravasation, dermal fibrosis, and adipocytic damage.

Differential diagnosis includes atrophic scarring

The differential diagnosis for hypopigmented atrophic macules and plaques over the lower limbs include atrophic scarring from previous trauma, guttate morphea, extra-genital lichen sclerosus, and tuberculoid leprosy.

Atrophic scarring occurs only after trauma.

Guttate morphea lesions are sclerotic and may be depressed.

Extra-genital lichen sclerosus is characterized by polygonal, shiny, ivory-white sclerotic lesions with or without follicular plugging.

Tuberculoid leprosy involves loss of nociception, hypotrichosis, and palpable thickened regional nerves (eg, great auricular, sural, or ulnar nerve).

Treatment requires long-term anticoagulation

Our patient had APS and the mainstay of treatment is long-term systemic anticoagulation along with attentive wound care.[6] Warfarin is preferred over a direct oral anticoagulant as it is more effective in the prevention of recurrent thrombosis in patients with APS.[7]

Our patient was started on warfarin. Since APS may occur as a primary condition or

in the setting of a systemic disease, such as systemic lupus erythematosus, she was referred to a rheumatologist. JFP

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