

Equestrian Perniosis

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Equestrian Perniosis

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Figure 1. Erythematous and violaceous scaling plaques on the left lateral thigh.



Figure 2. Erythematous and violaceous scaling plaques on the right lateral thigh.

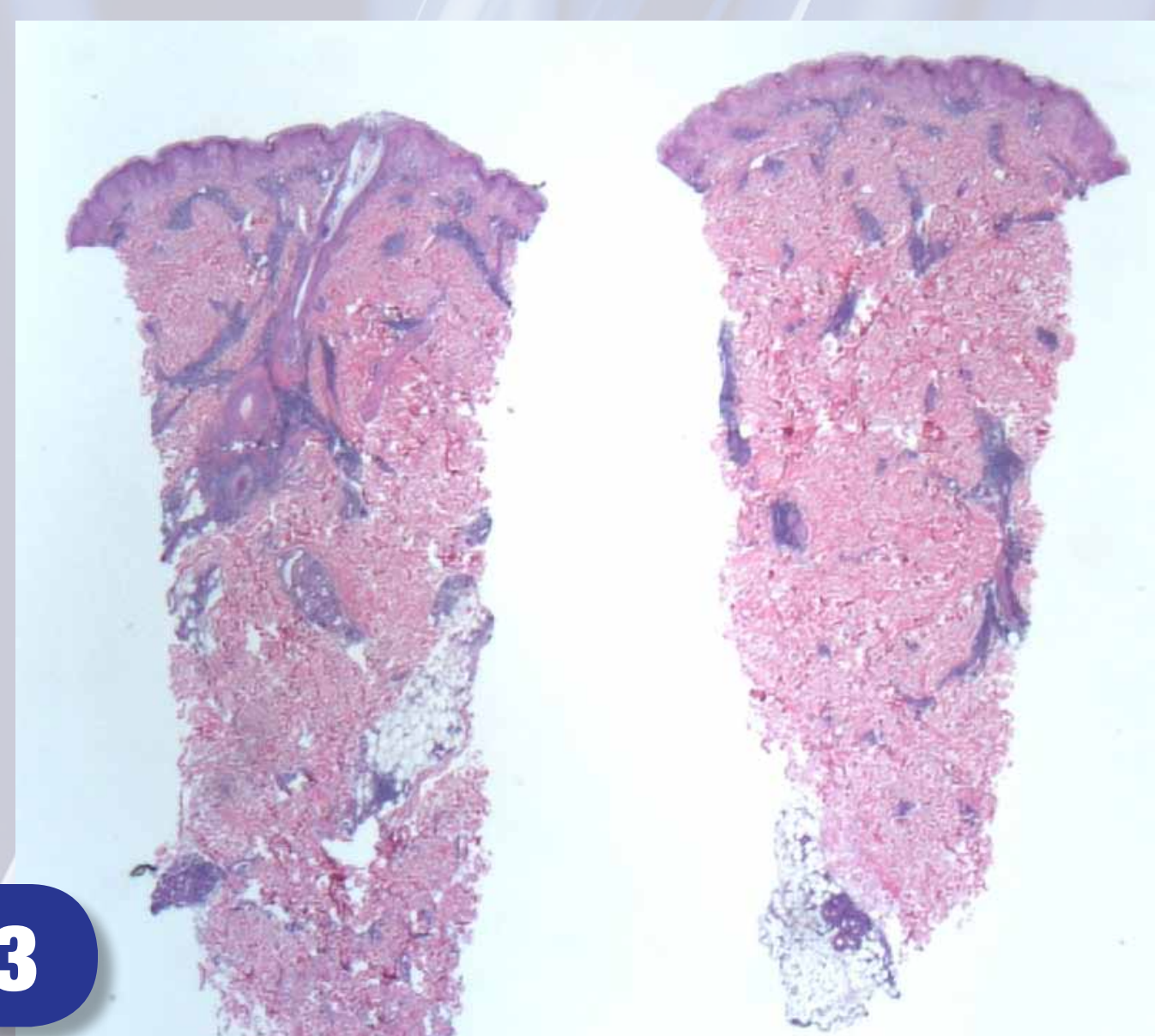


Figure 3. H&E (2X) Punch biopsy from the right thigh demonstrating a perivascular and periadnexal infiltrate.

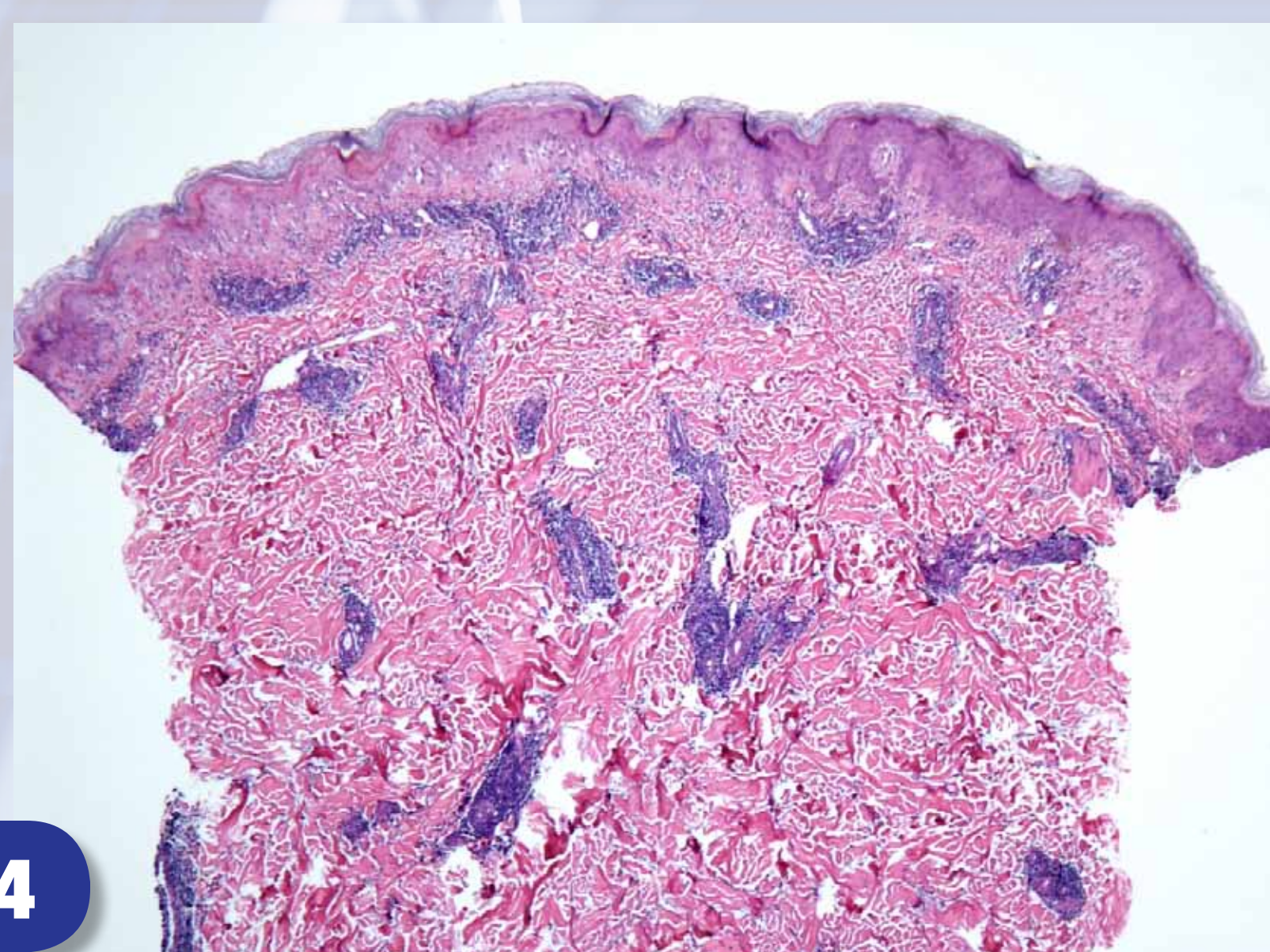


Figure 4. H&E (10X) Punch biopsy from the right thigh demonstrating a perivascular lymphocytic infiltrate, vacuolar interface changes, papillary dermal edema and dermal mucin.

Patient: S.H. is a 35 year-old male.

History of Present Illness: This patient presented with recurrent, painful and occasionally pruritic lesions on his lateral thighs and knees. The patient is an active equestrian and reports that the lesions appear in cold weather and resolve completely upon exposure to warm weather.

Medical History/Surgical History: None

Family History: Sister with lupus erythematosus

Previous Medications: Hydroxychloroquine and prednisone

Medications: None

Physical Examination: Multiple indurated pink to violaceous urticarial plaques on the lateral thighs and knees.

Laboratory Data: ANA, SSA/SSB, cold agglutinins, cryoglobulins, cryofibrinogens, CBC, CMP, ESR all WNL

Studies: None

Biopsy: Penn Cutaneous Pathology Services (1059986, 1/27/2011) Right and left lateral thigh: "interface reactions associated with superficial and deep dermal lymphocytic inflammation. There is an infiltrate comprised of mature lymphocytes throughout the dermis. This infiltrate exhibits a perivascular and periappendageal pattern. Prominent mucin deposition is seen in some areas. An interface reaction associated with vacuolar change and scattered necrotic keratinocytes was also noted. Mild epidermal hyperplasia and hyperkeratosis were seen. The infiltrate also extends into the subcutis and is associated with mucin production."

Reason for Presentation: Interest and treatment options

Discussion: Lesions of perniosis have been described as erythematous, tender nodules and plaques seen on the fingers, toes, heels, legs, thighs, nose and ears. In severe cases blistering and ulceration may occur. It often appears several hours after exposure to cold and resolves over one to three weeks. Lesions may become chronic especially in patients with vascular abnormalities. Perniosis is thought to be caused by intermittent or prolonged cold-induced vasoconstriction with the development of ischemia and vessel wall edema. They can appear as an idiopathic dermatosis or in association with an underlying autoimmune disease.

A specific type of perniosis has been observed in female horse riders in winter months with characteristic erythrocyanotic plaques distributed on the lateral thighs and buttocks. The reported lesions developed in cold and wind prone areas covered by tight fitting uninsulated riding pants after several hours of activity. Most patients have no known abnormal laboratory values, although the literature describes two patients with elevated cold agglutinins. Interestingly, the literature does not report a case of equestrian perniosis in a male, as in our case. It does however, report four male patients who developed tender pink to purple lesions of the lateral thighs consistent with perniosis after crossing a glacier-fed river. These men had limited protection of the thighs by shorts and experienced symptoms for a period of one week after exposure.

The pathogenesis of perniosis is unknown. The lateral thighs and buttocks are highly insulated with adipose tissue and because of the inherent decreased blood flow these areas may also experience a cold-induced increase in blood viscosity. The histological features observed include extensive dermal vessel edema with a superficial and deep perivascular lymphocytic infiltrate. Thickened small vessel walls and endothelial cell edema without fibrin deposition are seen.

Treatment of equestrian perniosis includes riding for shorter periods of time while wearing warmer, looser, and insulated undergarments. Peripheral arterial vasodilators may be effective in treatment and prevention. Nifedipine 20 mg three times a day improves circulation, reduces pain and resolution time, and prevents the development of new lesions. Avoidance of nicotine is also advised.

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