Late-Onset Focal Dermal Elastosis: A Case Report and Review of the Literature

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The estimated time to complete this activity is 1 hour.

GOAL

To understand late-onset focal dermal elastosis to better manage patients with the condition

LEARNING OBJECTIVES

Upon completion of this activity, you will be able to:

1. Recognize the clinical and histologic findings of late-onset focal dermal elastosis.
2. Differentiate late-onset focal dermal elastosis from pseudoxanthoma elasticum and linear focal elastosis.
3. Discuss theories of the pathogenesis of late-onset focal dermal elastosis.

INTENDED AUDIENCE

This CME activity is designed for dermatologists and general practitioners.

CME Test and Instructions on page 186.

This article has been peer reviewed and approved by Michael Fisher, MD, Professor of Medicine, Albert Einstein College of Medicine. Review date: March 2010.

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Late-onset focal dermal elastosis is a condition characterized by a localized increase in healthy-appearing elastic tissue in the mid and deep reticular dermis. The condition may clinically mimic pseudoxanthoma elasticum (PXE) and linear focal elastosis. We report a case of an 87-year-old woman who presented with a markedly thickened, yellow, pruritic plaque on the posterior neck and discuss the clinical and histopathologic distinctions between late-onset focal dermal elastosis, PXE, and linear focal elastosis.

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Case Report

An 87-year-old woman presented with a plaque of 3 months' duration on the left side of the posterior neck that had become increasingly pruritic. She

denied any pain or tenderness of the area. There was no history of gastric hemorrhage, stroke, or decreased visual acuity. Her medical history was unremarkable, except for hypothyroidism, hypertension, and hypercholesterolemia. There was no family history of similar skin lesions.

Physical examination revealed a 4.5×4.0-cm, thickened, yellow plaque with an erythematous border (Figure 1). A full-body examination did not reveal similar lesions in another location.

Results of a punch biopsy from the posterior neck showed actinic damage with an increased amount of grey solar elastosis in the reticular dermis. The papillary dermis was spared and contained pink collagen bundles without amorphous elastotic material (Figure 2). There was a marked increase in elastic fibers with Verhoeff elastic tissue stain and a focal increase in healthy-appearing elastic tissue in the mid and deep reticular dermis. There was no evidence of calcification or elastolytic change (Figure 3). A von Kossa stain did not show calcification of the fibers.

The patient was treated with mometasone furoate ointment 0.1% applied twice daily for several months. Noticeably decreased inflammation and erythema on the periphery of the lesion was achieved, and the pruritus was remarkably improved.

Comment

Late-onset focal dermal elastosis was first described in 2 elderly Japanese patients by Tajima et al[1] in 1995. Since then, few cases have been reported in the literature. The condition presents as firm, strikingly yellow papules that may coalesce into thickened plaques. The lesions typically are located on the neck or flexural surfaces of the arms and legs, with a predilection for the antecubital fossa and popliteal fossa. The lesions may be pruritic or entirely asymptomatic.

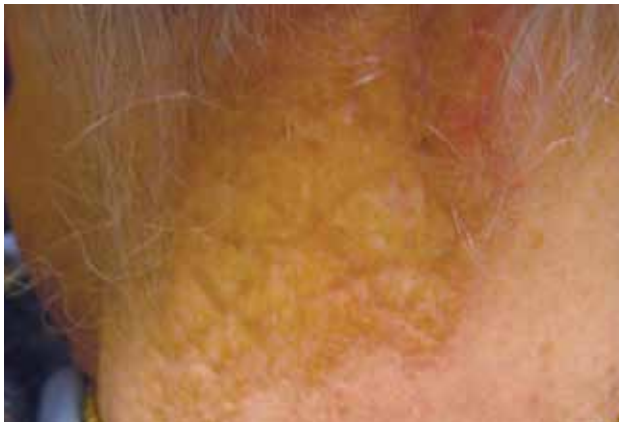


Figure 1. A thickened yellow plaque with an erythematous border on the left side of the posterior neck.

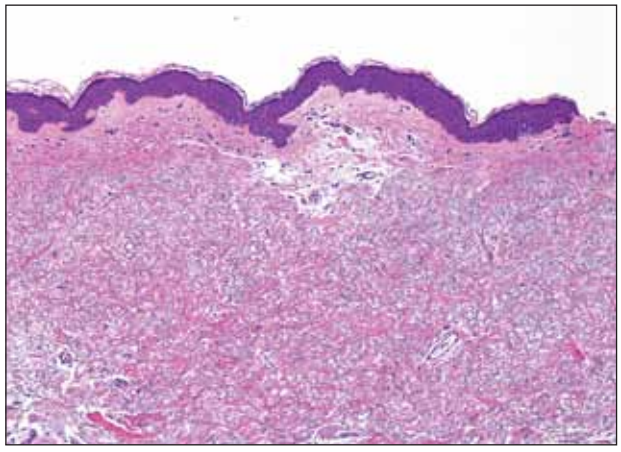
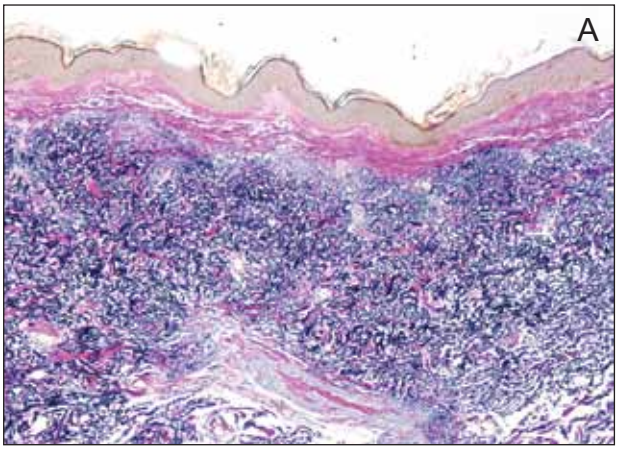


Figure 2. Results of a punch biopsy from the posterior neck showed actinic damage with an increased amount of grey solar elastosis in the reticular dermis. The papillary dermis was spared and contained pink collagen bundles without amorphous elastotic material (H&E, original magnification ×40).



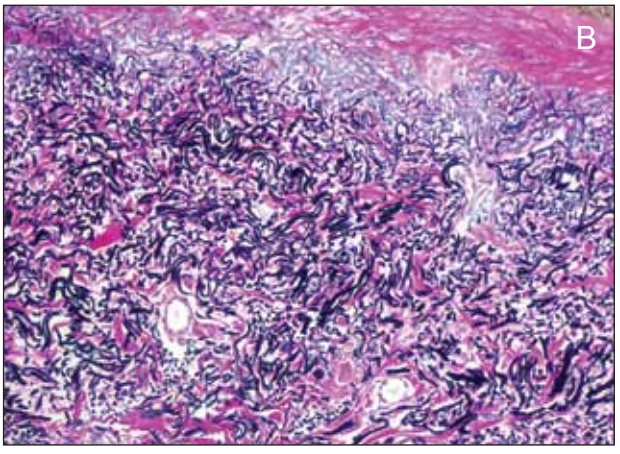


Figure 3. Results of a punch biopsy revealed a marked increase in elastic fibers (A) and a focal increase in healthy-appearing elastic tissue in the mid and deep reticular dermis (B)(Verhoeff elastic tissue; original magnifications ×40 and ×100, respectively). There was no evidence of calcification or elastolytic change.

The condition may clinically mimic pseudoxanthoma elasticum (PXE), but several important distinctions exist. Pseudoxanthoma elasticum is an inherited defect of elastic tissue characterized by yellow papules typically located on the sides of the neck and flexural areas.[2] Additionally, patients with PXE have varying degrees of systemic manifestations of their disease, including accelerated atherosclerosis, hypertension, cerebrovascular accidents, intermittent claudication, gastric hemorrhage, and ocular angioid streaks. Histologically, PXE is associated with granular, fragmented, curled, frayed, and thickened calcified elastic fibers in the mid and deep reticular dermis.[2]

Late-onset focal dermal elastosis has no associated systemic findings and histology shows a focal increase in healthy-appearing elastic tissue in the mid and deep reticular dermis. There is no evidence of calcification, elastolytic change, or extensive solar damage.[3]

Late-onset focal dermal elastosis also is clinically and histologically distinct from linear focal elastosis. Linear focal elastosis presents as striae-like, thickened, yellow lines on the lumbar region or, more rarely, the face or extremities. Linear focal elastosis preferentially affects males and is not an age-dependent process. Histologically, linear focal elastosis features elongated wavy elastic fibers with some fragmentation at the level of the mid dermis. The elastic fibers may be either thickened or thinned.[4]

The pathogenesis of late-onset focal dermal elastosis has not been determined, but several theories have been proposed. The usual absence of solar elastosis, the distinct histopathologic findings, and the presentation in typically sun-protected areas suggest that the condition is an intrinsic aging process rather than a condition secondary to sun damage. Tajima et al[1] reported that the disorder may result from increased elastin synthesis rather than a reduction in the elastic tissue degradation process. Kossard[5] postulated that the pathogenesis may be attributable to a local overproduction of structurally healthy tissue due to some loss of an age-related homeostatic growth regulating gene control mechanism.

The changes to elastic fibers are irreversible in late-onset focal dermal elastosis, and treatment is targeted to provide symptomatic relief of pruritus. High-potency topical corticosteroids, as successfully used in our patient, generally are effective. Although its pathogenesis has not been determined, late-onset focal dermal elastosis is a distinct clinical and histopathologic entity that may be underrecognized.

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