# cutis photo quiz

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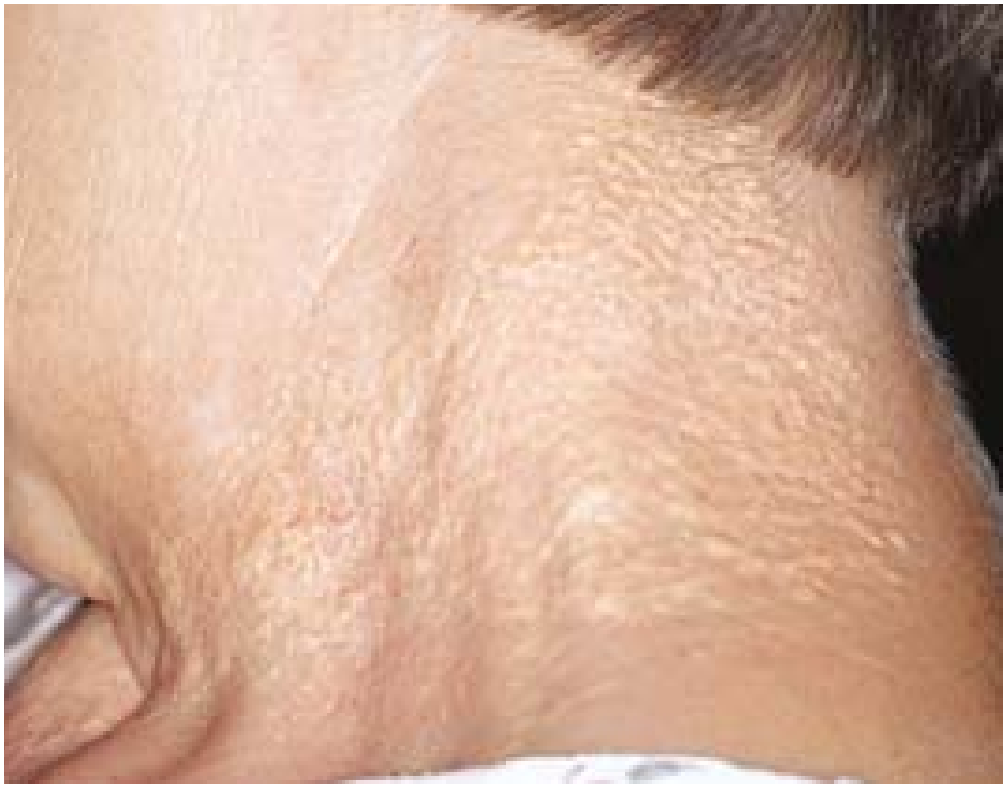

A 40-year-old woman presents with a plaque on her neck, suggesting a “gooseflesh” appearance.

## What is your diagnosis?

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# The Diagnosis

## Discussion

Pseudoxanthoma elasticum (PXE) is an uncommon, inherited disorder of elastic tissue that most frequently affects the skin, the retina, and arterial walls. Approximately 1 in 100,000 individuals are affected, with no gender predominance.[1] The disease is usually inherited in an autosomal recessive pattern, but autosomal dominant transmission also appears to occur.[2]

The pathogenesis of PXE remains unclear, but the primary pathophysiologic process is the calcification of elastic fibers in the dermis, the subretinal elastic layer (Bruch's membrane), and the elastic lamina of arteries. These changes account for the clinical presentation of PXE. The characteristic skin lesions are yellow patches or plaques of lax skin resembling peau d'orange or plucked chicken skin in flexural areas such as the neck, axilla, antecubital, popliteal, as well as the inguinal and periumbilical areas.[3] Biopsy of skin lesions is essential to establish the diagnosis. Histologic sections stained with hematoxylin and eosin will show mid-dermal, fragmented, and basophilic elastic fibers. A von Kossa stain may demonstrate calcification of these elastic fibers.

The retinal changes in PXE are referred to as angoid streaks, which are tears in Bruch's membrane just beneath the retina, and occur in roughly 85% of patients. They are seen on ophthalmologic examination as red-brown, curvilinear bands, radiating outward from the optic disk.[4] Arterial elastic calcifications occur in peripheral, coronary, and gastrointestinal tract vessels and result in claudication, hypertension, angina, and gastrointestinal hemorrhage. Once the diagnosis is made, first-degree relatives should also be screened for involvement. Appropriate specialty referrals would include dermatology, ophthalmology, and, if symptomatic, cardiology or gastroenterology.

## REFERENCES


1. Spitz JL: Genodermatoses. Williams & Wilkins, New York, 128-129, 1996.
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3. Hacker SM, Ramos-Caro FA, Beers BB, *et al.*: Juvenile pseudoxanthoma elasticum: recognition and management. *Ped Dermatol* 10: 19-25, 1993.
4. Fitzpatrick TB: Dermatology in General Medicine. McGraw-Hill, New York, 1843-1844, 1999.