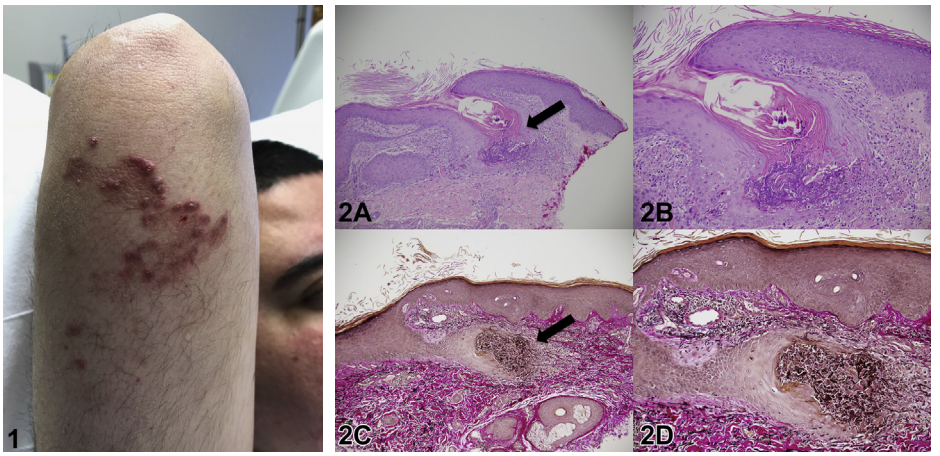
Keratotic papules in an annular arrangement

M. David Meyer, MD, and Sandra Osswald, MD
San Antonio, Texas

Key words: elastosis perforans serpiginosa; keratotic papule; perforating disorder; Verhoeff-van Gieson.



A 29-year-old man with history of hypertension and polymyositis presented with an asymptomatic rash to his upper arms that was present for more than a year. No medications were started before the appearance of the rash. He further denies any photosensitivity. Physical examination found erythematous, annular, keratotic papules and plaques localized to the upper posterior arms (Fig 1). An antinuclear antibody test result was negative. Results of a punch biopsy and special staining from one of the lesions are shown in Fig 2.

Question 1: What is the most likely diagnosis?

A. Pseudoxanthoma elasticum (PXE)

B. Granuloma annulare (GA)

C. Elastosis perforans serpiginosa (EPS)

D. Psoriasis

E. Papulosquamous subacute cutaneous lupus erythematosus (SCLE)

Answers:

A. PXE — Incorrect. Small, yellow papules and plaques, particularly on the neck, in addition to retinal changes are characteristics of PXE patients.

B. GA — Incorrect. GA lesions characteristically lack any scale or surface change as opposed to this patient's physical examination findings.

From the University of Texas Health at San Antonio.
Funding sources: None.
Conflicts of interest: None disclosed.
Correspondence to: M. David Meyer, MD, 7979 Wurzbach Rd, Mail Code 7876, Grossman Bldg, 3rd Floor, San Antonio, Texas 78229. E-mail: davidmeyer44@me.com.
JAAD Case Reports 2018;4:851-3.

2352-5126

https://doi.org/10.1016/j.jdcr.2018.06.020

C. EPS — Correct. The clinical features in conjunction with the histopathologic findings are strongly suggestive of EPS, a disorder more commonly seen in childhood or young adulthood, which is distinguished by the transepidermal elimination of abnormal elastic fibers. As seen in Fig 2, *A* and *B*, the most striking histopathologic finding is the presence of transepidermal channels that contain a combination of degenerated eosinophilic elastic fibers and basophilic debris. A chronic inflammatory infiltrate consisting of lymphocytes and histiocytes may also be present in the dermis. Occasionally, epidermal acanthosis and hyperkeratosis may be appreciated. Fig 2, *C* and *D* are discussed in Question 2.[1,2]

D. Psoriasis — Incorrect. The patient doesn't have any other stigmata of psoriasis, nor are the histopathologic findings consistent with psoriasis.

E. SCLE — Incorrect. The patient denies any photosensitivity, a common feature of SCLE. His antinuclear antibody test was also negative, making this entity less likely.

Question 2: What type of histochemical stain was performed to confirm the diagnosis?

A. Periodic acid—Schiff (PAS)

B. Fontana-Masson,

C. Von Kossa

D. Verhoeff-van Gieson (VVG)

E. Congo red

Answers:

A. PAS — Incorrect. PAS stain is useful for staining glycogen and elucidating fungal infections.

B. Fontana-Masson — Incorrect. Fontana-Masson stain is useful for evaluation of pigmentation disorders.

C. Von Kossa — Incorrect. Von Kossa is specific for calcium.

D. VVG — Correct. VVG is one of the most commonly used collagen and elastin stains. It stains elastic fibers black and collagen red. This stain is shown in Fig 2, *C* and *D*. The transepidermal, eosinophilic strands appreciated in the hematoxylin-eosin section (Fig 2, *A* and *B*) exhibit black staining with VVG (Fig 2, *C* and *D*), thus confirming the diagnosis of elastosis perforans serpiginosa.[1,2]

E. Congo red — Incorrect. Congo red stains amyloid with a green birefringence under polarized light.

Question 3: Which of the following is classically associated with the development of elastosis perforans serpiginosa (EPS)?

A. Scleredema

B. Cephalosporin administration

C. Bloom syndrome

D. Ehlers-Danlos syndrome

E. Bullous pemphigoid

Answers:

A. Scleredema — Incorrect. EPS is not associated with scleredema; however, it is associated with scleroderma.[2,3]

B. Cephalosporin administration — Incorrect. EPS has not been associated with cephalosporin administration, although it is associated with D-penicillamine use.[2,3]

C. Bloom syndrome — Incorrect. EPS is not associated with Bloom syndrome, but it has been reported to occur in patients with Rothmund-Thomson syndrome. Notably, both Bloom syndrome and Rothmund-Thomson syndrome occur because of mutations in DNA helicase.[2,3]

D. Ehlers-Danlos syndrome — Correct. Ehlers-Danlos syndrome has been linked to EPS development. Other associated conditions include Down syndrome, osteogenesis imperfecta, Marfan syndrome, acrogeria, Rothmund-Thomson syndrome, scleroderma, pseudoxanthoma elasticum, and cutis laxa. It is also seen in the setting of D-penicillamine administration.[2,3]

E. Bullous pemphigoid — Incorrect. Bullous pemphigoid and other related immunobullous conditions are not associated with EPS.[2,3]

Abbreviations used:

EPS: elastosis perforans serpiginosa
GA: granuloma annulare
PAS: periodic acid—Schiff
PXE: pseudoxanthoma elasticum
SCLE: subacute cutaneous lupus erythematosus
VVG: Verhoeff-van Gieson

REFERENCES

1. Lewis KG, Bercovitch L, Dill SW, Robinson-Bostom L. Acquired disorders of elastic tissue: part I. Increased elastic

tissue and solar elastotic syndromes. *J Am Acad Dermatol*. 2004;51(1):1-21.
2. Polańska A, Bowszyc-Dmochowska M, Żaba RW, Adamski Z, Reich A, Dańczak-Pazdrowska A. Elastosis perforans serpiginosa: a review of the literature and our own experience. *Postepy Dermatol Alergol*. 2016;33(5):392-395.
3. Mehta RK, Burrows NP, Payne CM, Mendelsohn SS, Pope FM, Rytina E. Elastosis perforans serpiginosa and associated disorders. *Clin Exp Dermatol*. 2001;26(6):521-524.