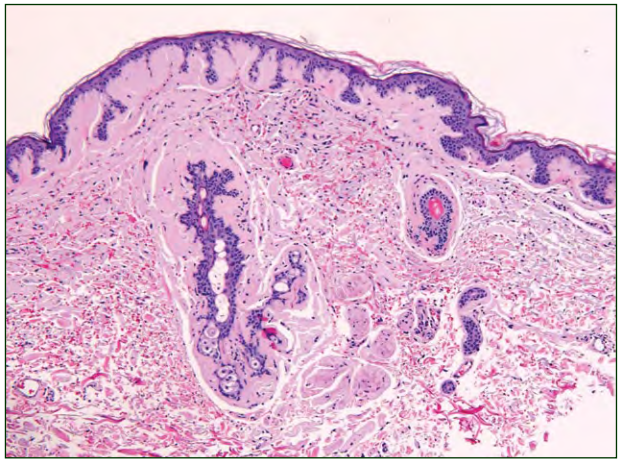
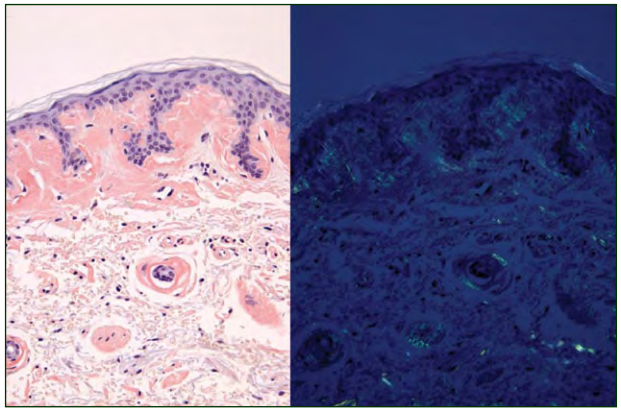


H&E, original magnification ×100.



Congo red, original magnification ×200 (left). Under polarized light, original magnification ×200 (right).

A biopsy was performed of a purpuric lesion on the left supraclavicular area of a 77-year-old woman. The best diagnosis is:

a. colloid milium
b. erythropoietic protoporphyria
c. lichen sclerosus
d. lipoid proteinosis
e. primary systemic amyloidosis

PLEASE TURN TO PAGE 123 FOR DERMATOPATHOLOGY DIAGNOSIS DISCUSSION

Suthinee Rutnin, MD; Rosalie Elenitsas, MD

Both from the Department of Dermatology, Hospital of the University of Pennsylvania, Philadelphia. Dr. Rutnin also is from the Department of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand.
The authors report no conflict of interest.
Correspondence: Suthinee Rutnin, MD, Division of Dermatology, Department of Medicine, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, 270 Rama VI Rd, Ratchatewi, Bangkok, Thailand 10400.

Primary Systemic Amyloidosis

Primary systemic amyloidosis, usually with an underlying plasma cell dyscrasia, is associated with a wide spectrum of organ involvement. Skin infiltration by amyloid presents as waxy, translucent, or purpuric papules, nodules, and plaques. Petechiae, purpura, and ecchymosis often are observed, especially on the eyelids, due to infiltration of vessel walls. Histopathology is characterized by amorphous material in the dermis and subcutaneous tissue. Amyloid often is found around adnexal structures and within blood vessel walls (Figure 1),[1-3] staining pink with hematoxylin and eosin and metachromatic with crystal violet and methyl violet. Amyloid stained by Congo red gives an apple green birefringence when viewed under polarized light (Figure 2).

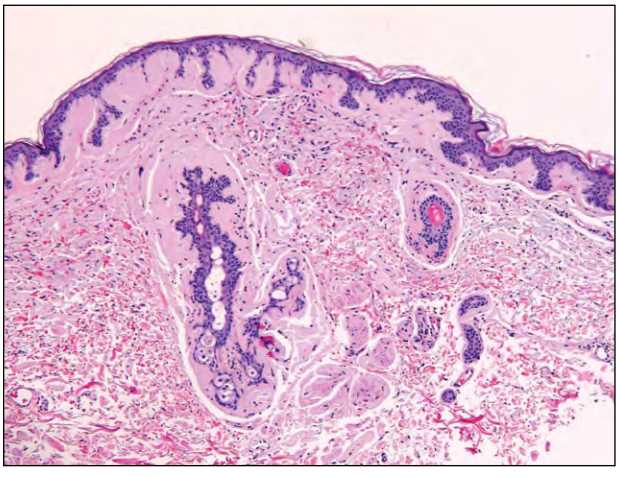


Figure 1. Amyloid of primary systemic amyloidosis (H&E, original magnification ×100).

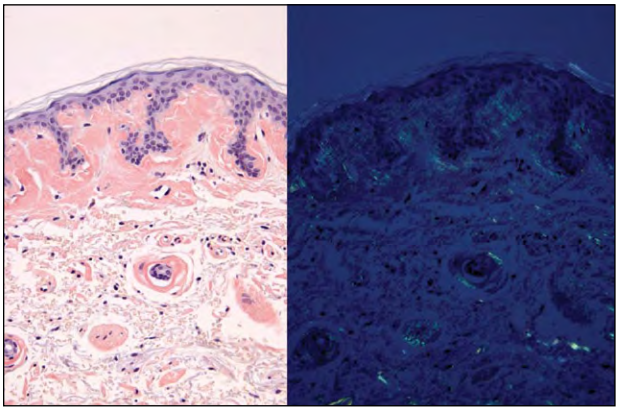


Figure 2. Amyloid of primary systemic amyloidosis staining positive with Congo red (left)(original magnification ×200) and producing an apple green birefringence when viewed under polarized light (right)(original magnification ×200).

Colloid milium stains similar to amyloidosis, except for pagoda red, which stains negative in colloid milium. Nodules of amorphous amphophilic materials are found in the papilla to mid dermis with artificial clefting and solar elastosis (Figure 3). Lipoid proteinosis is a progressive deposition of pale, eosinophilic, hyaline material in the superficial dermis. It surrounds the capillaries, which is known as an onionskin appearance, and is deposited in appendageal structures (Figure 4). The epidermis may show hyperkeratosis and some acanthosis in the verrucous lesion. This hyaline material contains

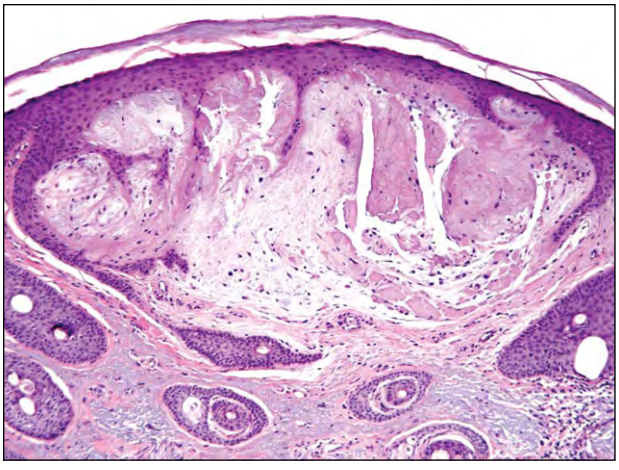


Figure 3. An amorphous amphophilic nodule with artificial clefting and solar elastosis characteristic of colloid milium (H&E, original magnification ×100).

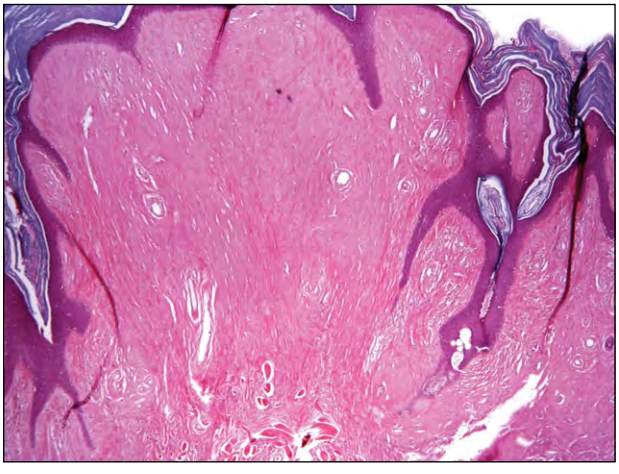


Figure 4. Amorphous eosinophilic material of lipoid proteinosis surrounding capillaries, known as an onionskin appearance, and eccrine sweat glands as well as in the thickened papillary dermis (H&E, original magnification ×40).

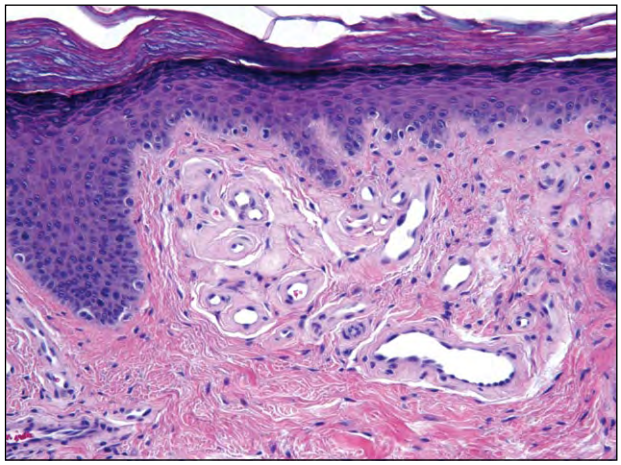


Figure 5. Hyaline material forming an irregular cuff around blood vessels in erythropoietic protoporphyria (H&E, original magnification ×200).

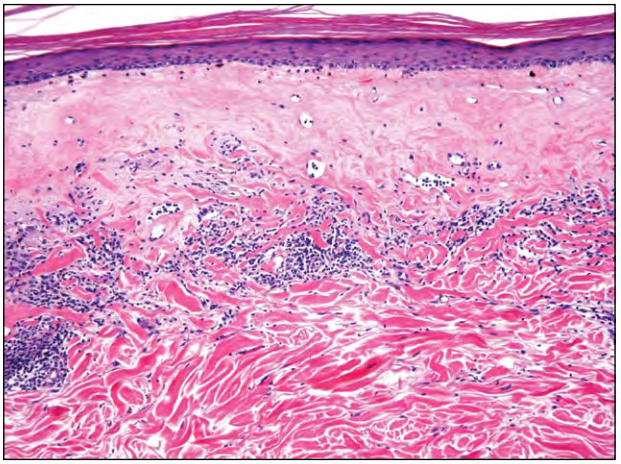


Figure 6. Homogenized collagen in the papillary dermis with thinning of the epidermis and vacuolar alteration of the basal layer characteristic of lichen sclerosus (H&E, original magnification ×100).

type IV collagen and laminin, which differs from amyloidosis by negative Congo red stain. In erythropoietic protoporphyria, the hyaline material not only involves the walls of small vessels in the papillary dermis but also forms an irregular cuff around these small vessels (Figure 5). However, it does not involve the sweat glands. In lichen sclerosus, homogenized collagen is situated above a diffuse perivascular infiltrate of lymphocytes, few plasma cells, and histiocytes. Epidermal changes include hyperkeratosis, follicular plugging, thinning of the epidermis, and vacuolar alteration of the basal layer (Figure 6).[4]

REFERENCES

1. Brownstein MH, Helwig EB. The cutaneous amyloidoses. II. systemic forms. *Arch Dermatol*. 1970;102:20-28.
2. Lee DD, Huang CY, Wong CK. Dermatopathologic findings in 20 cases of systemic amyloidosis. *Am J Dermatopathol*. 1998;20:438-442.
3. Touart DM, Sau P. Cutaneous deposition diseases. part I [published correction in *J Am Acad Dermatol*. 1998;39:1042]. *J Am Acad Dermatol*. 1998;39(2, pt 1):149-169.
4. Funaro D. Lichen sclerosus: a review and practical approach. *Dermatol Ther*. 2004;17:28-37.