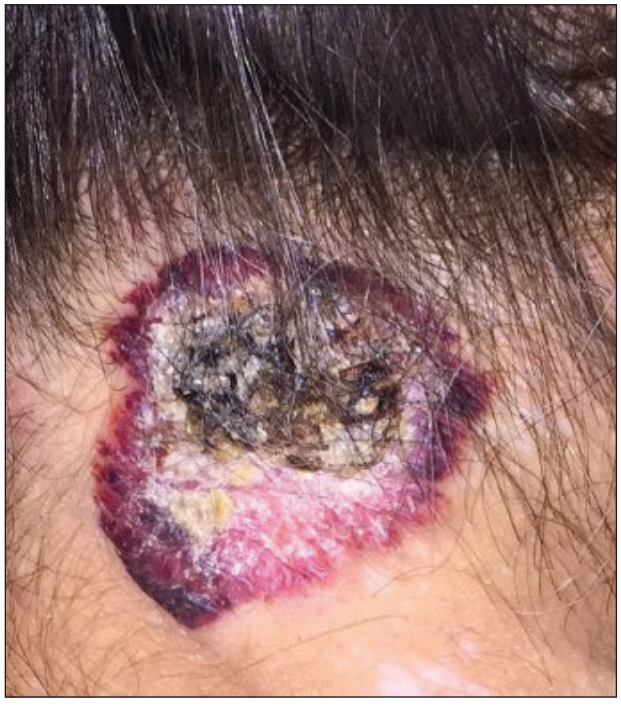
Large Hemorrhagic Plaque With Central Crusting

Caitlin Farmer, MD; Devika Patel, MD; Jason D. Pimentel, MD; Tor Shwayder, MD

Eligible for 1 MOC SA Credit From the ABD

This Photo Challenge in our print edition is eligible for 1 self-assessment credit for Maintenance of Certification from the American Board of Dermatology (ABD). After completing this activity, diplomates can visit the ABD website (http://www.abderm.org) to self-report the credits under the activity title "*Cutis* Photo Challenge." You may report the credit after each activity is completed or after accumulating multiple credits.



A 54-year-old woman with no notable medical history was referred to dermatology by her primary care provider for evaluation of a hematoma on the posterior neck that had developed gradually over 5 months. The lesion initially was asymptomatic but more recently had started to be painful and bleed intermittently. The patient denied any personal or family history of skin cancer. Physical examination revealed a large hemorrhagic plaque on the left side of the posterior neck with central brown-yellow crusting. There were few smaller, white, thin, sclerotic plaques with crinkling atrophy at the periphery of and inferolateral to the lesion. A punch biopsy specimen was obtained from the hemorrhagic plaque.

WHAT'S THE **DIAGNOSIS?**

a. angiosarcoma
b. bullous/hemorrhagic lichen sclerosus et atrophicus
c. ecthyma gangrenosum
d. lymphangioma
e. squamous cell carcinoma

PLEASE TURN TO PAGE 79 FOR THE DIAGNOSIS

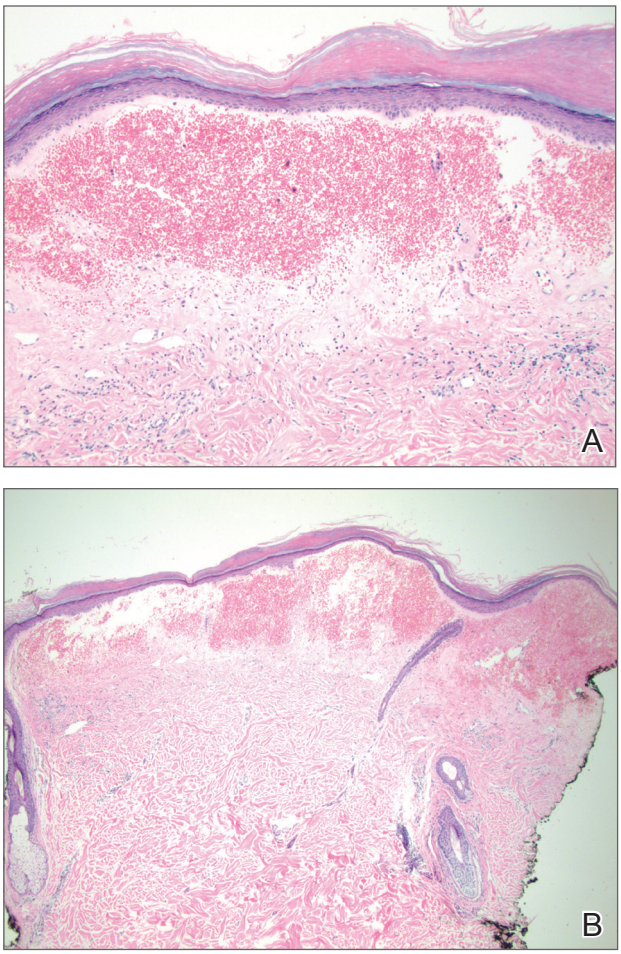
From the Department of Dermatology, Henry Ford Hospital, Detroit, Michigan.
The authors report no conflict of interest.
Correspondence: Caitlin Farmer, MD, 3031 W Grand Blvd, Ste 800, Detroit, MI 48202 (cfarmer2@hfhs.org).

THE **DIAGNOSIS:** Bullous/Hemorrhagic Lichen Sclerosus et Atrophicus

Histopathologic examination revealed hyperkeratosis of the stratum corneum and thinning of the epidermis (Figure). Subepidermal edema and hemorrhage in the papillary dermis were seen. There were dilated vessels beneath the edema in the reticular dermis, as well as perivascular, perifollicular, and interstitial lymphocytic inflammation. No cytologic atypia characteristic of squamous cell carcinoma (SCC) and angiosarcoma or large lymphatic channels characteristic of lymphangioma were noted. Based on clinicopathologic correlation, the diagnosis of the bullous/hemorrhagic form of lichen sclerosus et atrophicus (LS&A) was made. The patient was treated with high-potency topical steroids with notable symptomatic improvement and rapid resolution of the hemorrhagic lesion.

Lichen sclerosus et atrophicus is a chronic inflammatory condition with a predilection for the anogenital region, though rare cases of extragenital involvement have been reported. It is seen in both sexes and across all age groups, with notably higher prevalence in females in the fifth and sixth decades of life.[1,2] Lichen sclerosus et atrophicus can be difficult to diagnose, as these patients may present to a variety of specialists, may be embarrassed by the condition and reluctant for full evaluation, or may have asymptomatic lesions.[2,3] Rare cases of isolated extragenital involvement and hemorrhagic or bullous lesions further complicate the diagnosis.[1,2] Despite these difficulties, diagnosis is essential, as there is potential for cosmetically and functionally detrimental scarring as well as atrophy and development of overlying malignancies. Lichen sclerosus et atrophicus is not curable and rarely remits spontaneously, but appropriate treatment strategies can help control the symptoms of the condition as well as its most devastating sequelae.[3]

For females, classic LS&A is most common in the prepubertal, perimenopausal, or postmenopausal periods, commonly involving the vulva or perineum. Symptoms include pruritus, burning sensation, dysuria, dyspareunia, and labial stenosis, among others. For males, most cases involve the glans penis in prepubertal boys or middle-aged men, and symptoms include pruritus, new-onset phimosis, decreased sensation, painful erections, dysuria, and urinary obstruction.[1-3] An estimated 97% of patients have some form of genital involvement with only 2.5% showing isolated extragenital involvement, though the latter may be underdiagnosed, as this area is more likely to be asymptomatic.[3-6] Extragenital LS&A most often involves the neck and shoulders. The classic appearance of LS&A includes shiny, white-red macules and papules that ultimately coalesce into atrophic plaques and can be accompanied by fissuring or scarring, especially in the genital area.[2] There is an increased risk for SCC associated with genital LS&A.[1]



A, Histologic analysis showed hyperkeratosis of the stratum corneum, papillary dermal sclerosis and edema, zone of lymphoid inflammation, and copious erythrocyte extravasation (H&E, original magnification ×100). B, Copious erythrocyte extravasation also was evident in the superficial dermis (H&E, original magnification ×40).

Bullous/hemorrhagic LS&A has been described as a rare phenotype. One case report cited an increased incidence of this subtype in patients with exclusively extragenital lesions, and the authors considered blister formation to be a characteristic feature of extragenital LS&A. The pathogenesis of blister formation and hemorrhage in LS&A is not completely understood, but trauma is thought to play a role due to decreased stress tolerance from atrophic skin.[4] Furthermore, distortion of blood vessel architecture in LS&A has been described with loss of the capillary network and enlargement of vessels along

the dermoepidermal junction, which also could play a role in hemorrhage. Differential diagnosis of the bullous/hemorrhagic type of LS&A includes bullous pemphigoid, bullous lichen planus, or bullous scleroderma.[7] In our more exophytic hemorrhagic case, malignancies such as SCC or angiosarcoma also had to be considered. Unlike genital LS&A, extragenital LS&A including the bullous/hemorrhagic variant has not been linked to an increased risk for malignancy.[1,5]

The mainstay of treatment of all forms of LS&A is high-potency topical steroids, but topical retinoids, tacrolimus, and UVA phototherapy also have been used. Bullous/hemorrhagic lesions often resolve quickly with topical steroids, leaving behind more classic plaques in their place, which can be more refractory to treatment.[5,7]

REFERENCES

1. Meffert JJ, Davis BM, Grimwood RE. Lichen sclerosus. *J Am Acad Dermatol.* 1995;32:393-416.
2. Pugliese JM, Morey AF, Peterson AC. Lichen sclerosus: review of the literature and current recommendations for management. *J Urol.* 2007;178:2268-2276.
3. Fistarol SK, Itin PH. Diagnosis and treatment of lichen sclerosus: an update. *Am J Clin Dermatol.* 2013;14:27-47.
4. Kimura A, Kambe N, Satoh T, et al. Follicular keratosis and bullous formation are typical signs of extragenital lichen sclerosus. *J Dermatol.* 2011;38:834-836.
5. Khatu S, Vasani R. Isolated, localised extragenital bullous lichen sclerosus et atrophicus: a rare entity. *Indian J Dermatol.* 2013;58:409.
6. Luzar B, Neil SM, Calonje E. Angiokeratoma-like changes in extragenital and genital lichen sclerosus. *J Cutan Pathol.* 2009;36:540-542.
7. Lima RS, Maquine GA, Schettini AP, et al. Bullous and hemorrhagic lichen sclerosus—case report. *An Bras Dermatol.* 2015;90 (3 suppl 1):118-120.