

Verrucous papule on thigh

Was this lesion the result of sun exposure, or a recent trauma to the patient's leg?

A 21-year-old man came into our medical center to have a lesion on his thigh examined. He said the lesion developed a few months earlier at the site of minimal trauma. He noted that, over the previous few months, the lesion had progressively darkened and it bled sporadically. On examination, we noted a solitary 7.5-mm firm, blue-black verrucous papule over the right medial thigh (FIGURES 1A AND 1B). There

were no other lesions.

The patient indicated that he had gotten sunburned many times in the past. He also said that he had an aunt who'd had a melanoma.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

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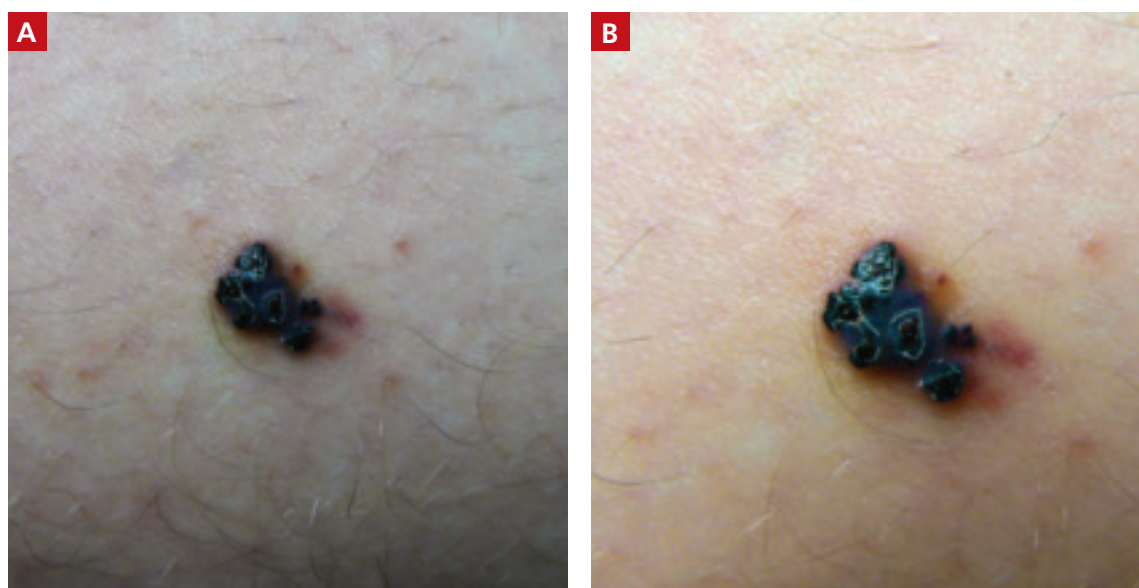
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FIGURE 1

A lesion that bled sporadically



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This papule on the medial aspect of the patient's right thigh developed after minimal trauma to the area. The lesion had been getting progressively darker.

Diagnosis: Angiokeratoma

An angiokeratoma is a benign pink-red to blue-black variably sized papule or plaque that is typically 2 to 10 mm in diameter.¹ Angiokeratomas are composed of a series of subepidermal dilated capillaries that have a characteristic hyperkeratotic surface and bleed easily.² These lesions are rare, with a prevalence estimated to be 0.16% in the general population.³

The pathogenesis of angiokeratoma formation is unclear; however, multiple theories exist. The development of these lesions may be related to repeated trauma or friction at a particular site.⁴ Alternatively, increased venous blood pressure or primary degeneration of vascular elastic tissue could explain their development.⁵ While their cause is unclear, the initial event in the development of an angiokeratoma is believed to be the development of a vascular ectasia within the papillary dermis. The epidermal reaction appears to be a secondary phenomenon due to increased proliferative capacity on the surface of the vessels.⁵

■ **The most common form**—as seen in this case—is the solitary or sporadic angio-

keratoma. It comprises 70% to 83% of all cases of angiokeratomas³ and usually develops on the lower extremities. Angiokeratomas typically arise during the first 2 decades of life,⁶ and are more common in men.³ Other types of angiokeratomas include angiokeratoma of Mibelli, angiokeratoma of Fordyce, angiokeratoma circumscriptum, and angiokeratoma corporis diffusum (Fabry's disease).^{7,8}

■ **Angiokeratoma of Mibelli** is characterized by pink to dark red papules or verrucoid nodules that occur most commonly in men⁷ and involve the bony prominences, such as the elbows.

■ **Fordyce lesions** involve the scrotum or vulva and are usually numerous and related to conditions with elevated venous pressure.

■ **Angiokeratoma circumscriptum** usually present as papules that commonly coalesce to form plaques.

■ **Fabry's disease**, or angiokeratoma corporis diffusum, is an X-linked recessive disease related to a deficiency in alpha-galactosidase A. This leads to multiple, variably sized angiokeratomas occurring in childhood that are concentrated between the umbilicus and the knees. This disease invari-

> **The development of these lesions may be related to repeated trauma or friction at a particular site.**

FIGURE 2
A view from the dermatoscope

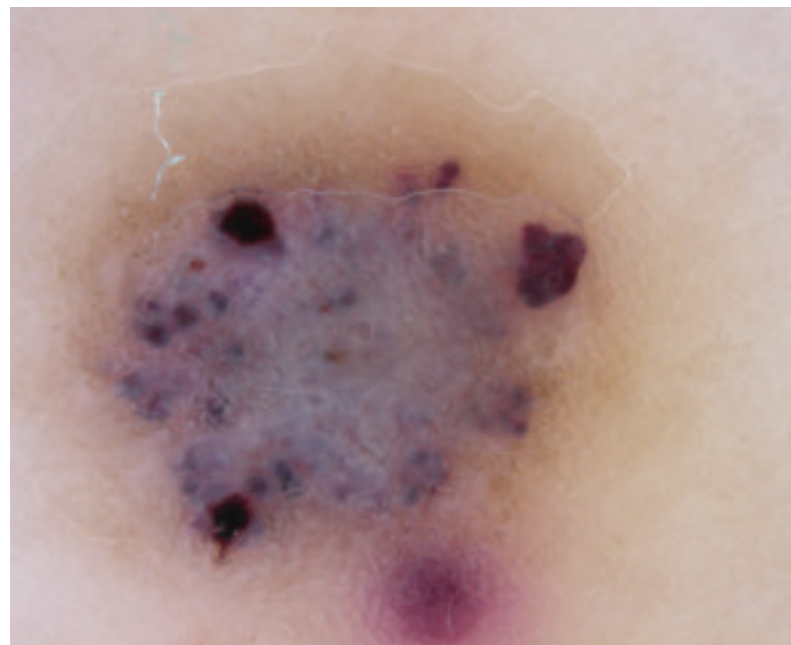


PHOTO COURTESY OF RICHARD P. USATINE, MD

This image reveals the purple lacunae that are characteristic of an angiokeratoma.

Nonmelanocytic melanoma mimickers

A number of nonmelanocytic lesions can be confused with melanoma. They include the following:

■ **Actinic keratoses (AKs)** are a type of keratinocytic neoplasm that typically develops on the sun-exposed skin of the elderly. An AK is typically 3 to 10 mm in size, pink to red in color, and has scaling secondary to local hyperkeratosis. If these lesions are left untreated, they can develop into squamous cell carcinomas (SCCs) at a rate of 0.24% annually.^{15,16} Thus, AKs are more often a concern for SCC than for melanoma. However, the pigmented variant of an AK can clinically and histologically raise concern for melanoma due to its pigmentation and microscopic evidence of melanin within keratinocytes and macrophages.¹⁵ If it is not possible to differentiate an AK from melanoma clinically or histologically, immunohistochemistry is often required to make the final diagnosis. For example, immunohistochemical staining with S-100 can be used to identify epidermal melanocytes and distinguish them from atypical keratinocytes.¹⁷

■ **Basal cell carcinoma (BCC)** is the most common skin cancer.¹⁸ While most BCCs are amelanocytic, 7% of BCCs are pigmented and present as irregularly pigmented nodules with irregular telangiectatic vessels on their surface. The center of a BCC may be depressed or ulcerated and may easily crust or bleed. Definitive diagnosis may be made histologically. A BCC typically consists of columns of basaloid cells with atypical nuclei, sparse cytoplasm, and peripheral cellular palisading.¹⁹ BCCs are easily differentiated from melanoma using immunohistochemistry, as they are negative for traditional melanocytic markers.¹⁷

■ **Seborrheic keratoses (SKs)** are among the most common skin lesions and represent a benign proliferation of immature keratinocytes. The appearance of an SK can vary from a smooth peppered appearance to a rough surface that may be irregularly pigmented, dry, and fissured. Given their range of presentation, it is common for SKs to be biopsied to evaluate for melanoma and occasionally BCC.²⁰

■ **Dermatofibromas (DFs)** are common benign skin lesions that typically appear as pink-to brown-colored firm nodules that represent a localized response to skin injury and inflammation. DFs are typically 3 to 10 mm in diameter and are most commonly located on the anterior surface of the thigh. Histologic analysis of a DF reveals an acanthotic epidermis with a proliferation of spindle cells in the mid and lower dermis, with capillaries dispersed throughout. A common finding in DFs is the trapping of collagen within the spindle cell at the periphery of the lesion.²¹

➤ **When the clinical diagnosis is in doubt, the entire lesion should be removed with narrow margins.**

ably leads to involvement of other organs, which may result in renal failure, myocardial infarction, or cerebrovascular accidents.^{1,7}

A mimicker of melanoma

An angiokeratoma is an uncommon, though important, mimicker of melanoma. (For more on other lesions that can be confused with melanoma, see “Nonmelanocytic melanoma mimickers” above.)

Melanoma is the most aggressive and potentially life-threatening neoplasm in the differential diagnosis of an angiokeratoma. Risk factors for melanoma include increasing age, fair skin and hair color, tendency for freckling, number of moles (5 large or >50 small nevi doubles the risk of melanoma), a personal or first-degree family history of melanoma, and a history of intermittent sunburns.⁹⁻¹²

CONTINUED

How to diagnose angiokeratoma

The clinical presentation typically suffices in making the diagnosis of an angiokeratoma. If dermoscopy is performed, the characteristic findings include the presence of scale and purple lacunae¹³ (FIGURE 2). However, when there is suspicion of melanoma or the clinical diagnosis is in doubt, the entire lesion should be removed (with narrow margins) in order to obtain a definitive diagnosis. Histological findings consist of dilated subepidermal vessels associated with epidermal hyperkeratosis.³

No need to treat, unless there are cosmetic concerns

If the diagnosis is straightforward and a biopsy is not needed, no treatment is necessary because simple angiokeratomas are

benign entities. However, treatment may be considered for cosmetic purposes, or to prevent bothersome bleeding. Angiokeratomas may be removed via shave or standard excision, electrodesiccation and curettage, or destroyed with a laser. For Fabry's disease, in which numerous angiokeratomas pose a cosmetic concern, laser therapy, including the use of an argon, copper, Nd:Yag, KTP 532-nm, or Candela V-beam laser, is preferred.¹⁴

■ **In our patient's case**, we performed a 2-mm punch biopsy, which revealed that the lesion was an angiokeratoma. It was subsequently removed by shave biopsy with clear margins. **JFP**

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➤ Treatment may be considered to prevent bothersome bleeding.

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