PHOTO ROUNDS

Dusky plaque on the knee

Our patient wasn't surprised that she got a scar after a recent fall. But she was surprised when it started to grow

progressively growing lesion on the left knee prompted a 35-yearold woman to visit our clinic. She reported that about 3 months earlier, she had developed a small ulceration on the knee following a fall. With local wound care, the ulceration healed with a scar. The scar, however, continued to grow and she developed distinct papules outside the original scar.

The scar subsequently became raised with violaceous discoloration. The patient reported having no history of excessive scarring or keloid forming after skin surgery or trauma. There was neither a personal nor family history of inflammatory or infectious granulomatous diseases.

On physical examination, there was

an erythematous to dusky plaque with well-defined irregular borders. There were also discrete papules on the anterior and medial aspect of the knee. The plaque measured approximately 2.8 cm by 3.8 cm. There were no tender nodules on the shins, nor was lymphadenopathy present. A routine chest x-ray was normal.

To support our clinical diagnosis, we took a 4-mm punch biopsy from the center of the plaque. The histologic examination revealed changes in the dermis termed noncaseating "naked" granulomas.

What is your diagnosis?

How would you manage this condition?

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Histologic exam revealed noncaseating "naked" granulomas

Scar develops into a dusky plaque



A scar on this 35-year-old woman's left knee continued to grow to a 2.8 cm by 3.8 cm dusky plaque with well-defined, irregular borders and discrete papules on the anterior and medial aspect of the knee.

FIGURE

Diagnosis: Scar sarcoidosis

Sarcoidosis is a systemic granulomatous disease that may affect any organ system, and therefore may present with various clinical manifestations.¹ Sarcoidosis can be an incidental finding on chest x-ray or be discovered in patients that present with respiratory or constitutional symptoms.²

Cutaneous sarcoidosis occurs in up to one third of patients with systemic sarcoidosis.² The classic skin lesions are "erythema nodosum"—an acute, nodular, erythematous eruption that usually is limited to the shins, and "lupus pernio" red-to-purple or violaceous indurated nodules affecting the nose, cheeks, ears, and lips. There are other uncommon skin presentations of sarcoidosis ranging from scattered papules and annular lesions to erythrodermic skin manifestations.³

In scar sarcoidosis, there is spontaneous development of livid or reddishbrown plaques on scars that were previously atrophic for the most part. Scar sarcoidosis may be caused by:⁴⁻⁶

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"Naked" granuloma refers to the absence of surrounding lymphocytes seen in scar sarcoidosis

- venipuncture
- tuberculin skin tests
- herpes zoster
- tattoos
- cosmetic fillers such as hyaluronic acid injection.

Infection and other factors may be at work

Although the precise cause of sarcoidosis remains unknown, various infectious, noninfectious, environmental, and genetic factors may be at work. Researchers have theorized that immune dysregulation may be involved. Contact with a persistent antigen that is poorly cleared by the immune system may lead to T lymphocytes and mononuclear phagocytes accumulating in the granulomas of sarcoidosis.⁷ Researchers have proposed that inoculation of foreign matter from minor trauma may be one type of pathogenic mechanism in cutaneous sarcoid.⁸

Granuloma annulare is part of the differential

A wide range of diseases comprise the differential diagnosis of sarcoidosis. These diseases include:⁹

• Granuloma annulare. It is also a granulomatous skin disease, but it appears as single or multiple rings.

• **Rheumatoid nodules.** These usually appear in the context of a diagnosis of rheumatoid arthritis.

• Granulomatous mycosis fungoides. This type of cutaneous lymphoma has many clinical forms, including granuloma formation.

• **Syringoma.** On inspection, you'll see small, firm adnexal benign tumors that usually appear around the upper cheeks and lower eyelids

• Xanthelasma. These are benign, yellow macules, papules, or plaques that tend to appear on the eyelids. Patients with xanthelasma often have a lipid disorder.

• Lichen planus. This is a very pruritic skin involvement with pink to violaceous papules and plaques. It may present in different locations, but the most common areas are the wrists and ankles.

• **Granulomatous rosacea**. This is a variant of rosacea characterized by uniform papules on the face.

Clinical findings, biopsy clinch the diagnosis

The diagnosis of sarcoidosis is made by a combination of clinical and histologic findings.¹

Clinical findings. Cutaneous involvement is either "specific" or "nonspecific."

• With specific cutaneous involvement, which our patient had, you'll see typical noncaseating granulomas, with no evidence of infection or a foreign body. It may be disfiguring, but it's almost always nontender and it is rarely ulcerative.

• With nonspecific cutaneous involvement, you'll see erythema nodosum lesions, especially on the legs. The serum angiotensin-converting enzyme (ACE) level is elevated in many of these patients. **Histologic findings.** Skin biopsy demonstrating noncaseating granulomas provides definitive evidence of skin involvement. Typical sarcoid lesions are characterized by circumscribed granulomas of epithelioid cells with little or no necrosis. (The term "naked" granuloma refers to the absence, or small number, of surrounding lymphocytes.)

Other granulomatous diseases, such as berylliosis and tuberculosis, must be excluded since they often present the same way as scar sarcoidosis.⁷

Steroids control symptoms, slow disease progression

Topical, intralesional, and systemic corticosteroids are used to treat scar sarcoidosis, as are systemic medications such as chloroquine¹⁰ and allopurinol.¹¹ Corticosteroids (local and systemic) are effective in controlling all sarcoid symptoms; they also slow disease progression.¹

For localized skin involvement, intralesional corticosteroids are typically more effective than topical steroids. Systemic corticosteroids are reserved for widespread, progressive lesions or those that impair function.^{1,12,13} A starting dose of 1 mg/kg of prednisone is appropriate.

In general, the prognosis of cutaneous sarcoidosis is good.² The course is variable, ranging from self-limited acute episodes to a chronic debilitating disease that may result in death.² Spontaneous remissions occur in nearly two thirds of patients, but 10% to 30% have a more chronic or progressive course.^{1,2,13}

Our patient responds to treatment

Our patient declined intralesional corticosteroid injections, so we started her on potent topical corticosteroid tapes (Cordran). She had significant improvement 6 weeks later.

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Disclosure

The authors reported no potential conflict of interest relevant to this article.

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Topical, intralesional, and systemic corticosteroids are used to treat scar sarcoidosis

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