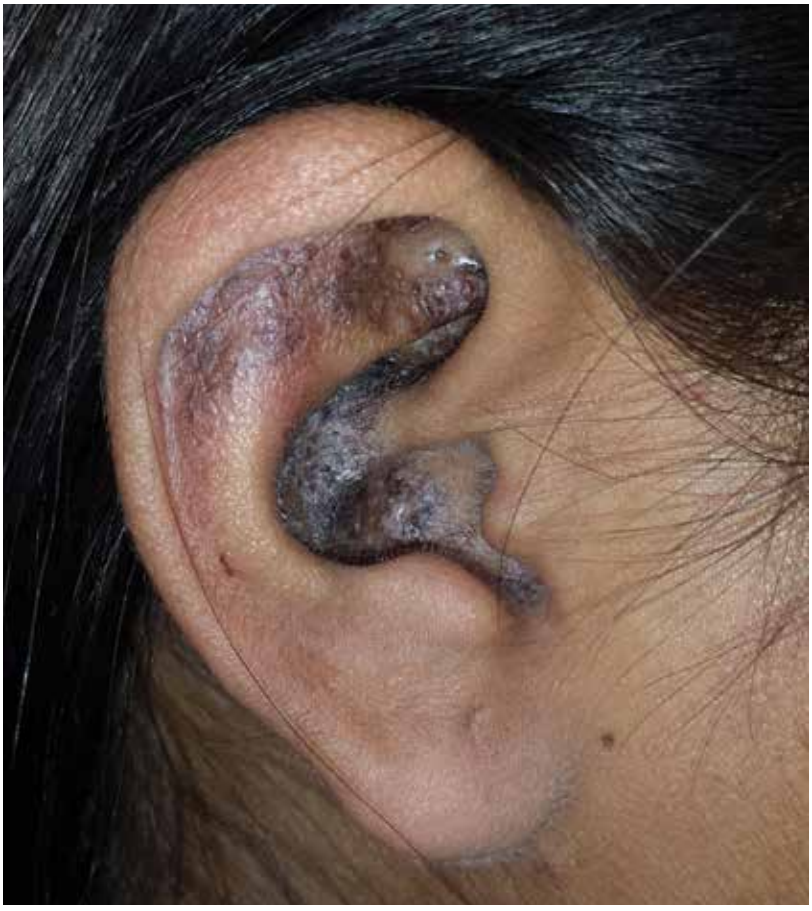
The Ears Have It (and She Doesn't Want It)

A 34-year-old black woman is sent to dermatology by her rheumatologist for evaluation of changes to her ears that began several months ago. The patient reports no symptoms, but she is quite distressed by the appearance of her ears.

She has been under the care of the rheumatologist for several years for her systemic lupus erythematosus. She takes hydroxychloroquine (400 mg/d), which she says controls most of her systemic symptoms (ie, joint pain and malaise).

Further history taking reveals that, within the time frame of the ear changes, the patient also developed an itchy rash on both arms. Application of triamcinolone 0.1% cream has not helped.

On examination, the changes to the patient's ears are immediately obvious: the dark brown to black discoloration contrasts sharply with her light brown skin. In addition to the color change, the surface of the ears is scaly and rough, with enlarged pores evident. There is no redness or swelling noted; palpation elicits neither increased warmth nor adenopathy around the ears or on the adjacent neck.



The scaly rash on the patient's arms is remarkably symmetrical. It affects the sun-exposed lateral portions of both arms, sparing the skin on the medial aspects and on the proximal portions normally covered by clothing.

Of the following, the best next step to take in this case is

a) Increase the patient's hydroxychloroquine dose to 400 mg bid
b) Start the patient on an oral antibiotic (eg, cephalexin 500 mg qid)
c) Start a two-week taper of prednisone 60 mg
d) Obtain a biopsy to establish a firm diagnosis, if possible

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ANSWER

The correct answer is to obtain a biopsy (choice "d"), the results of which would likely dictate rational and effective treatment. The other choices are largely empirical and not based on available evidence.

DISCUSSION

In this case, the biopsy (with samples from the arm rash as well as from the ears) showed unequivo-

cal evidence of connective tissue disease—almost certainly lupus.

Systemic lupus erythematosus (SLE) has protean manifestations because it can affect so many different organs in so many different ways. Reduced to its simplest elements, lupus is an autoimmune process that results in a form of vasculitis that can affect any perfused tissue.

In terms of the skin, the visible manifestations of lupus are numerous and not always obvious. Sun is a known exacerbating factor, as this case demonstrated quite well: The patient's rash was pronounced on sun-exposed skin but spared covered skin. When this was brought to her attention, the patient recalled a baby-sitting job earlier in the year (summer) that had required her to spend time outdoors. She also acknowledged that her smoking habit often took her into the backyard, where she would stand in the sun.

That being said, neither the arm rash nor the ear changes are "typical" of lupus (although the latter did include *patulous follicular orifices,* enlarged pores often seen focally with lupus). The effect is simply the result of the patient's normal dark skin color; on a white person, the discoloration would have been pink or red.

Although these changes were suspicious for lupus, it was necessary to establish the diagnosis by biopsy—especially since the patient was already being treated for the disease. With that accomplished, the patient was sent back to her rheumatologist, who indicated he would probably treat her with a biologic, plus or minus methotrexate. **CR**