CASE 1

A 62-year-old woman is concerned about a rapidly enlarging lesion on her right shoulder that she first noticed six months ago. Her medical history is notable for hypertension, diabetes, and moderate sun exposure. There is no personal or family history of skin cancer.

The lesion is 1.6 cm wide, erythematous, and unaccompanied by pain, pruritus, or bleeding. The patient’s axillary nodes are nonpalpable. A biopsy is performed.

What is your diagnosis?

CASE 2

A 22-year-old woman presents to your emergency department with a chronic, erythematous, occasionally pruritic rash on her neck and torso. It has waxed and waned over the past seven years and is currently flaring. She notes that stress and sunlight exposure tend to exacerbate the condition, with symptoms worsening in late spring. Her mother and a maternal aunt are similarly afflicted.

You observe erythematous papules with a yellowish tint on her neck and upper torso. In addition, her fingernails are brittle and dystrophic.

What is your diagnosis?
The patient has Darier disease, also known as keratosis follicularis or Darier-White disease. This autosomal-dominant disorder is characterized by crusted, somewhat oily papules ranging in color from flesh-toned to sallow-hued. Common symptoms include pruritus, pain, and a foul odor. Heat and exposure to ultraviolet-B light exacerbate the rash. Nails may exhibit longitudinal ridges and splitting. The papules most commonly occur on the upper trunk and seborrheic areas of the scalp and face. The standard treatment of sunscreen, emollients, and topical steroids in conjunction with oral retinoids, such as isotretinoin, may not completely control this difficult condition.

The patient’s lesion is a dermatofibrosarcoma protuberans (DFSP), a rare soft-tissue malignancy that is locally aggressive. Early diagnosis of DFSP is uncommon due to its non-descriptive clinical appearance and relatively slow growth (this case is atypical). This neoplasm is most commonly found on the trunk and proximal extremities. Due to the likelihood of local recurrence, the treatment of choice is wide excision, possibly followed by radiation therapy. Approximately 4% of DFSPs metastasize to the regional lymph nodes or lungs, which portends a grave prognosis.