

When the Scales Align



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Several years ago, this 50-year-old woman developed a rash under her breasts. It comes and goes, but in hot weather, the condition is particularly uncomfortable and accompanied by an objectionable odor.

The rash was declared a yeast infection by a previous provider, but it failed to respond to several different prescription and OTC anti-yeast creams, lotions, and oral medications. The patient is in no distress but is somewhat agitated about the lack of effective treatment. Referral to dermatology is arranged.

History-taking reveals a family history of skin problems. However, neither the patient nor any family member has ever seen a dermatologist before now, and no provider has ever biopsied the affected skin.

On examination, the florid, white, scaly rash under the patient's breast stands out in stark contrast to her type V skin. The rash is bilateral, and the affected area precisely aligns with the inframammary fold of each breast. There are sharp margins and uni-

form moist scaling.

No other areas of skin are affected. However, seven of her 10 fingernails exhibit longitudinal white and red streaks, and several nails have triangular nicks in the edges. The roof of her mouth is studded with fleshy nodules measuring 0.6 to 1 cm. Several pits are seen on her palms.

A sample of the affected inframammary skin is taken via punch biopsy and submitted to pathology. The report shows acantholysis with focal dyskeratotic keratinocytes. Intraepidermal separation is seen throughout the specimen.

Given these findings, the most likely diagnosis is:

- Yeast infection
- Psoriasis
- Intertrigo
- Darier disease

ANSWER

The correct diagnosis is Darier disease (choice "d"), also known as *Darier-White disease* or *keratosis follicularis*.

The outlined differential is reasonable when you consider (1) how unusual Darier's is and (2) how unlikely it is to manifest *solely* in the inframammary area. One must bear in mind that, as with psoriasis (choice "b") and seborrhea in this area, intertrigo (choice "c") is not always a primary process. And while yeast infection (choice "a") can complicate any florid rash in this area, the patient had tried topical and oral anti-yeast products with no success.

DISCUSSION

Darier disease is an inherited defect transmitted by autosomal dominant mode. It is relatively rare, occurring in 1:30,000 to 1:100,000 population, depending on the geographic area. The pathophysiologic process is a breakdown of cell adhesion that normally binds keratin filaments to tiny connecting fibers called *desmosomes*.

Darier's usually manifests in the third decade of life, with a "branny" papulosquamous

rash affecting the chest, scalp, back, and intertriginous areas. Men and women are equally affected, but the condition is more commonly seen in persons with darker skin.

When the condition is florid, it emits a distinct foul odor. Itching can be severe. The nail and intraoral findings seen in this patient are typical. In my experience, nail problems are the more common of the two and are essentially pathognomic for Darier disease.

TREATMENT

In cases such as this one, isotretinoin is used, though it will offer only temporary relief. Other options—oral antibiotics (eg, minocycline) and/or topical steroids (used with caution, given the risk for atrophy in this particular area)—are often sufficient for less severe cases.

At best, this patient's prognosis is guarded. But control of her condition is certainly possible. **CR**