REPORT OF CASES

CASE 1

A 22-year-old woman presented with a 2-year history of an eruption on the anterior aspect of her neck and right arm that was relatively asymptomatic. She had a history of cystinuria, which had been treated with D-penicillamine for several years, but the D-penicillamine therapy had been discontinued 2 years before the onset of the eruption. A biopsy was performed approximately 1 year before her presentation to our institution. The biopsy specimen demonstrated clawlike downgrowths of epidermis surrounding collections of amorphous basophilic debris and hyperplastic elastic fibers. Many elastic fibers were noted to be pushing through epidermal channels (Figure 1), a finding that was consistent with the clinical diagnosis of elastosis perforans serpiginosa (EPS). The patient was treated unsuccessfully with several modalities in a sequential fashion, including liquid nitrogen cryotherapy monthly for 6 months, topical tretinoin gel nightly for 2 months, oral isotretinoin at dosages ranging from 40 to 60 mg/d for 15 weeks, and 2 sessions of carbon dioxide laser surgery. On physical examination, she was noted to have erythematous annular and arcuate keratotic plaques on the anterior aspect of her neck and right arm (Figure 2).

CASE 2

A 56-year-old woman was referred to our institution for evaluation of a pruritic “nonhealing scar” that had been present on the posterolateral aspect of the left side of her neck for 1 year. The lesions developed 1 month after a revised facial rhytidectomy scar revision. The patient had been treated with oral cephalexin for 2 weeks and topical erythromycin solution for several weeks, without improvement. Her medical history revealed that she had been treated with D-penicillamine for Wilson disease for more than 20 years. The D-penicillamine therapy had been discontinued 2 months before her presentation to our clinic.

Physical examination revealed multiple 2- to 7-mm crusted, erythematous, ulcerated papules and plaques with an arcuate configuration. These lesions were associated with a 6-cm-long scar on the posterolateral aspect of the left side of the neck.

A biopsy specimen demonstrated a central focus of pseudoepitheliomatous hyperplasia that appeared to be...
connected to the epidermis by a channel of epithelium. Adjacent inflammation with dermal necrosis, both within and surrounding the epithelia, was evident. An elastic stain demonstrated an increased concentration of elastic fibers in the middermis extending to the epidermis. Many of the elastic fibers appeared clumped and thickened.

The patient was treated unsuccessfully with cryotherapy approximately 6 times; high-potency topical corticosteroids, including clobetasol and halobetasol, for 6 weeks; topical 0.1% tretinoin cream and topical 0.05% tretinoin solution for about 2 months each; and several intralesional injections of triamcinolone acetonide (4 mg/mL).

THERAPEUTIC CHALLENGE

Multiple therapies have been reported to be effective in the management of EPS. However, none has been universally accepted as the treatment of choice. Reported effective treatments include liquid nitrogen cryotherapy1 and oral isotretinoin therapy.2 Our patients were previously treated with many modalities, without success; therefore, topical tazarotene therapy was initiated.

SOLUTION

Because multiple therapies had failed in both cases, the patients were offered a trial of 0.1% tazarotene gel. Both patients agreed and began using 0.1% tazarotene gel at bedtime. At the 1-month follow-up visits, their disease was somewhat improved. After 2 months of tazarotene therapy, the condition of patient 1 was greatly improved (Figure 3) and that of patient 2 was moderately improved. After 4 more weeks, patient 2 was almost free of active disease. Patient 2 then discontinued tazarotene therapy, and her disease flared. Other topical retinoid preparations were then tried, without improvement. Cryotherapy was tried a few more times, also without improvement, and the patient was finally re-treated with tazarotene, which flattened her lesions within 6 weeks. Patient 1 has tried to taper her usage of tazarotene but notices flares on discontinuation.

The only adverse effect observed in both cases was mild irritation. However, the irritation subsided after a few weeks of therapy. Both patients continue to use tazarotene daily for intermittent courses when their disease flares.

COMMENT

Elastosis perforans serpiginosa is a disorder in which altered elastic fibers are recognized as foreign material and are extruded through the epidermis by transepidermal elimination. The result is a papular eruption that is usually arranged serpiginously, annularly, or arcuately. Many conditions are associated with EPS, including Down syndrome, Rothmund-Thomson syndrome, Ehler-Danlos syndrome, Marfan syndrome, osteogenesis imperfecta, and pseudoxanthoma elasticum. Also, patients treated with penicillamine are prone to develop EPS.3 Elastosis perforans serpiginosa usually occurs in young adults and shows a predilection for the head and neck.

Tazarotene is the first receptor-selective topical retinoid approved for the treatment of plaque psoriasis. It selectively targets the γ and β subtypes of retinoic acid receptors. Ninety percent of retinoid receptors in the skin are of the γ subtype. Hofmann et al4 reported tazarotene’s effectiveness in the treatment of congenital ichthyoses in an open, intraindividually controlled, half-side investigation. Burkhart and Burkhart5 reported tazarotene’s effec-

Figure 2. Patient 1 on presentation to our institution with erythematous annular plaques on her neck.

Figure 3. Patient 1 after 2 months of treatment with 0.1% tazarotene gel.
tiveness in treating a patient with Darier disease who had responded poorly to other agents. One mechanism of action of tazarotene in psoriasis is thought to be attributable to the down-regulation of keratins 6, 10, and 16. Tazarotene also has a strong antiproliferative effect via the expression of 3 genes: tazarotene-induced genes 1 through 3. Tazarotene has a low systemic absorption and is rapidly metabolized and eliminated. The most common adverse effect reported is local irritation. To our knowledge, this is the first report of EPS being successfully treated with tazarotene. The mechanism of action of tazarotene in treating EPS is unknown. Tazarotene may have comedolytic properties that allow for the unplugging of transepidermal pores in this disease. Also, the blockage of retinoic acid receptors may play a role in decreasing the proliferation in EPS.

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REFERENCES


Clinicians, local and regional societies, residents, and fellows are invited to submit cases of challenges in management and therapeutics to this section. Cases should follow the established pattern. Submit 4 double-spaced copies of the manuscript with right margins nonjustified and 4 sets of the illustrations. Photomicrographs and illustrations must be clear and submitted as positive color transparencies (35-mm slides) or black-and-white prints. Do not submit color prints unless accompanied by original transparencies. Material should be accompanied by the required copyright transfer statement, as noted in “Instructions for Authors.” Material for this section should be submitted to George J. Hruza, MD, Laser and Dermatologic Surgery Center Inc, 14377 Woodlake Dr, Suite 111, St Louis, MO 63017. Reprints are not available.