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CORRESPONDENCE

Efficacy of topical adapalene monotherapy for symptomatic relief in a long-standing vulvar syringoma: A case report and literature review with treatment update

Dear Editor,

Vulvar syringoma is a benign adnexal skin tumor of eccrine duct origin, localizing to the female genital area.[1] It is the second-most clinical variant following periorbital disease and often accompanies intractable pruritus,[2] representing a disease morbidity and treatment dilemma. We report such a case of vulvar and facial syringomas, whose persisted vulvar pruritis unresponsive to topical corticosteroids improved with topical adapalene monotherapy.

An otherwise healthy 53-year-old Japanese woman presented with a 10-year history of papules on her vulva with severe pruritis. Repeated topical corticosteroids were ineffective. Examination revealed miliary-sized, yellowish-white, multiple papules localized to the bilateral labia majora (Figure 1A,B). Similar eruptions were distributed on her periorbital region and cheeks (Figure 1C). She had neither family history nor experience of exacerbation during menstruation. Histopathology of biopsied vulvar skin revealed numerous small ducts in the fibrous dermal stroma (Figure 1D). The duct walls consisted of row epithelial (cuboidal) cells, which were lined by cuticle cells in the luminal inside (Figure 1E). Some of the ducts possessed comma-shaped tails of surrounding epithelial cells, so-called "tadpole" appearance (Figure 1F). This clinicopathology suggests facial and vulvar syringomas.

Considering the inefficacy of preceded topical corticosteroids, we challenged a topical adapalene gel of 0.1% (Differin® Gel 0.1%). Within 4-weeks of adapalene use, intractable vulvar pruritus dramatically faded with decreased swelling. By 3-months, the vulval papules became relatively smaller in size, some of which disappeared (Figure 1G,H). There were no adverse events. During 6-months of follow-up, the vulvar lesions remained a favorable symptomatic control, albeit without much difference in appearance.

Vulvar syringoma typically appears in puberty of females and tends to become worse during pregnancy or with the use of oral contraceptives, implicating a hormonal imbalance. Conservative treatments may be applicable because topical corticosteroid remains a value of choice as initial treatment and spontaneous regression may occur. The English literature search found 118 cases with syringoma at least appeared in the vulvar area.[3–5] Of these, 46 cases (38.9%) had pruritus, requiring some sort of anti-pruritic treatment. A half (59/118, 50%) received any of the treatments, most frequently with topical corticosteroids (30/59, 50.8%), which were mostly unhelpful (20/30, 66.7%).

Adapalene is a low-molecular-weight naphthoic acid derivative (MW 412.5) with retinoid-like activity and is widely approved for acne treatment. The literature search found only four cases, including ours, with syringomas at least affecting the vulva and whose skin lesions responded to either adapalene or all-trans retinoid tretinoin.[6,7] All cases had a previous history of topical corticosteroid that was unhelpful but achieved flattening and/or size reduction of individual papules. Our case obtained a significant improvement of intractable pruritis within a few weeks of adapalene initiation.

Adapalene harbors variable pharmacological actions, including anti-inflammatory effects via inhibition of proinflammatory cytokines released from the corresponding keratinocytes and inflammatory infiltrates, and increase of extracellular matrix synthesis with a decrease of matrix metalloproteinase activity in dermal fibroblasts expressing adapalene-bound retinoic acid receptor-β.[8,9] One may consider that adapalene stimulates the turnover and remodeling of dermal matrix/appendages, together with anti-inflammatory and anti-comedogenic actions,[10] contributing to the favorable clinical efficacy in our case.

DECLARATION SECTION

Approval of the research protocol: No human participant was involved in this study.

Informed Consent: Written informed consent was obtained from the patients.

Registry and the Registration No. of the study/trial: N/A.

Animal Studies: N/A.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

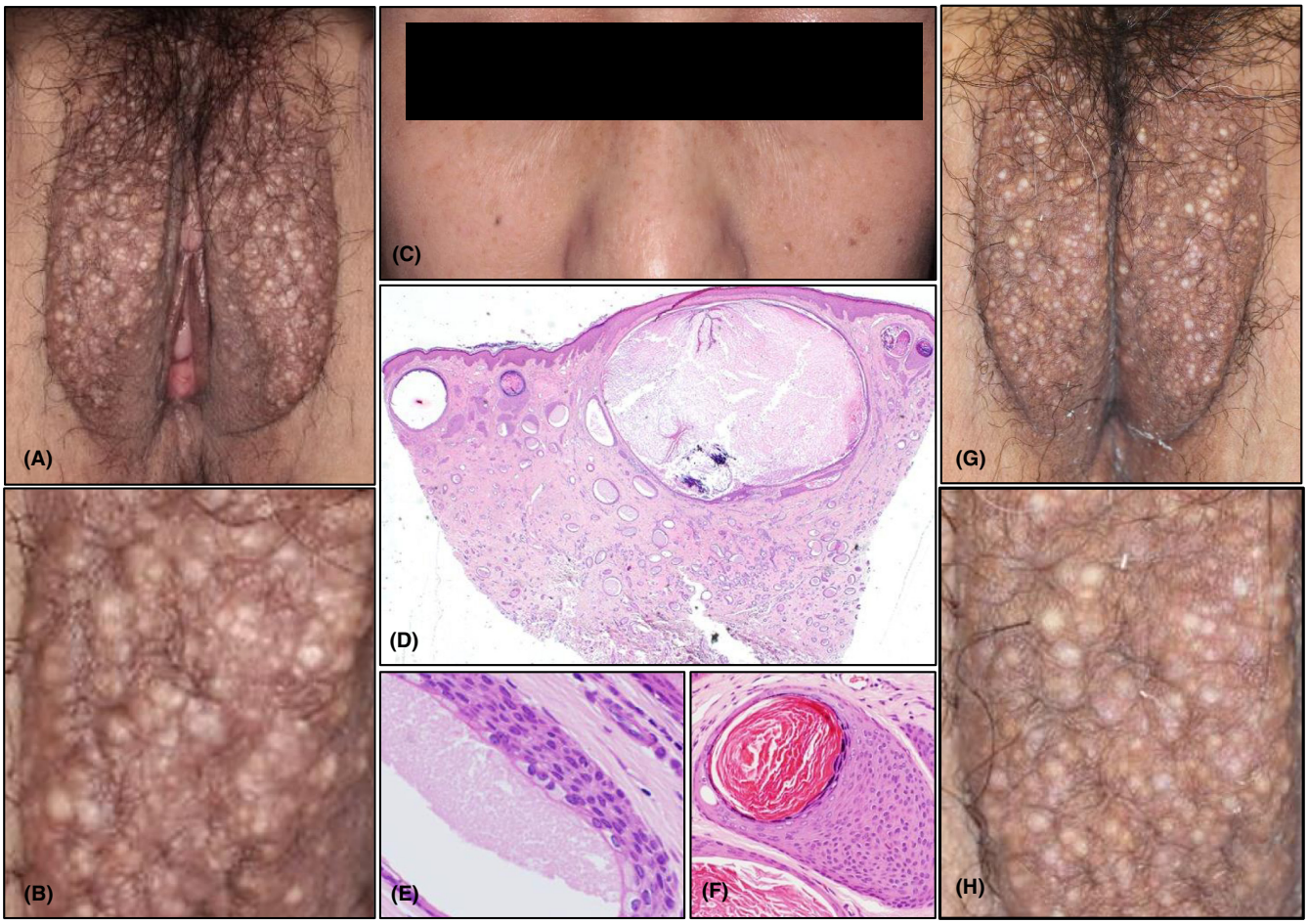

FIGURE 1 (A) There were miliary-sized, yellowish-white, multiple papules localizing to the bilateral labia majora of the vulva. (B) High-power view of the pretreated vulvar lesion. (C) Similar whitish papules scattering on the periorbital region and cheeks. (D) Histopathology of the vulvar skin showing numerous small ductal structures in the fibrous dermal stroma (×40, HE staining). (E, F) The duct walls consisting of row epithelial cells, lined by cuticle cells in the luminal inside. Some of the ducts possessed comma-shaped tails of epithelial cells, consistent with tadpole appearance (×200, HE staining). (G) By 3-months of topical adapalene use, the vulval papules became smaller in size with decreased swelling of the bilateral labia majora, some of which disappeared. (H) High-power view of the vulvar lesion after 3-months of adapalene treatment

Saori Uesugi-Uchida MD[1]
Noritaka Oyama MD, PhD[1]
Yoshio Yoshida MD[2]
Minoru Hasegawa MD, PhD[1]

[1]*Department of Dermatology, Division of Medical Sciences, University of Fukui, Fukui, Japan*
[2]*Department of Gynecology, Division of Medical Sciences, University of Fukui, Fukui, Japan*

Correspondence
Noritaka Oyama, MD, PhD, Department of Dermatology, Faculty of Medical Sciences, University of Fukui, 23-3 Matsuoka-Shimoaizuki, Eiheiji, Fukui 910-1193, Japan.
Email: norider@u-fukui.ac.jp

ORCID
Saori Uesugi-Uchida https://orcid.org/0000-0001-7192-0128

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