Dowling–Degos Disease Affecting the Vulva

Sir,

In 1974, Wilson-Jones & Grice (1) reported cases of reticular pigmented macules of the flexures demonstrating a unique epidermal naevoid anomaly. They clearly showed that the syndrome was not to be confused with any type of acanthosis nigricans. Earlier case reports by Dowling & Freudenthal (2) and Degos & Ossipowski (3) provided the foundation for this new entity, which is inherited by an autosomal dominant gene.

There have only been a few previous case reports of Dowling–Degos disease (DDD) of the vulva (1, 4, 5). Their skin lesions are usually confined to the external genital areas, with a characteristic histology showing filiform downgrowth of pigmented epithelial strands 2–4 cells wide, extending from the interfollicular epidermis and follicular infundibulum. No report has come from Asia. We describe here the case of a Japanese female affected by DDD of the vulva.

CASE REPORT

A 55-year-old Japanese female had been treated for pruritus vulvae for 5 years by a local gynaecologist, who later noticed the presence of innumerable, small, pigmented macules on her bilateral vulva. She stated that none of her family members had similar eruptions.

The eruptions were deep brownish-black pigmented spots less than 1 mm in diameter, which were distributed symmetrically only on the bilateral external genital regions (Fig. 1).

A biopsy specimen showed irregular, filiform epidermal elongations of hyperpigmented rete ridges with a concentration of melanin at the tips (Fig. 2). These changes also extended from the follicular infundibulum. The filiform epidermal downgrowths closely resembled those of senile lentigo or adenoid seborrhoeic keratosis. Hyperpigmentation was present in the digitations as finely dispersed melanin granules scattered uniformly throughout the cytoplasm of the cells, the pigment not being deposited as supranuclear caps as in normal skin. There was no atypicality in keratinocytes or melanocytes.

DISCUSSION

The genital pigmented spots observed in this patient showed the typical clinical and histopathological features of DDD. There was a lack of other affected family members and no involvement in other locations, which initially caused diagnostic difficulty. DDD must be distinguished from acanthosis nigricans, adenoid seborrhoeic keratosis, senile lentigo and other hereditary pigmented anomalies. Disregarding the follicular changes, there was close similarity to adenoid seborrhoeic keratosis.

Topical application of adapalene, a naphtholic acid derivative with retinoid-like pharmacology, was reported to be a successful treatment for a female patient with an overt pruritic form of DDD (6). In other reports hydroquinone, tretinoin, topical steroids and systemic retinoids have been tried without success (7). Our patient did not want any treatment for this condition.

Fig. 1. Close-up view of the pigmentary eruptions in a 55-year-old Japanese female. The deep brownish-black pigmented spots were less than 1 mm in diameter, and distributed symmetrically on the bilateral external genital regions.

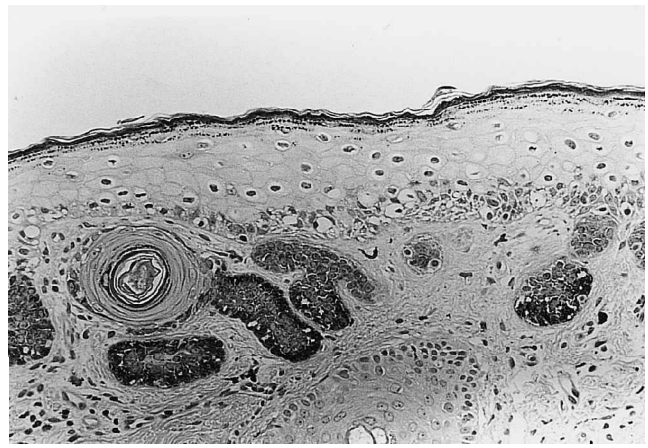

Fig. 2. Photomicrograph of the histopathology of the small pigmented spots. Filiform epidermal downgrowth of the hyperpigmented rete ridges was found in the interfollicular spaces.

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