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CPD

Superimposed segmental dermatosis papulosa nigra

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Dermatosis papulosa nigra (DPN) is a common condition in individuals with Fitzpatrick skin phototypes IV–VI.[1] There is a familial aggregation.[2] In black populations, the prevalence of DPN ranges between 35 and 77%, whereas the frequency is lower but still high in indigenous Central and South American or Asian populations.[3] Hence, it is obvious that DPN has a polygenic background. Most authors agree that DPN should be categorized as a variant of seborrhoeic keratosis (SK), and molecular studies appear to support this view.[4] We report a case of unilateral linear DPN of the face associated with bilateral ordinary facial lesions of the same disorder.

A 32-year-old woman of indigenous Bolivian origin presented with a curvilinear arrangement of pigmented papules involving the right side of her face, running from the temporal area to the mandible (Fig. 1) in a Blaschko-linear distribution. She reported that some of these lesions had probably been present at birth, and she was certain that they had existed during early infancy. She asked for advice on how to treat these cosmetically disturbing skin changes. In addition to the linear lesions, some nonsegmental papules of similar appearance were noted on the patient's right ear, right preauricular area, below the right corner of the mouth, the right side of her lower lip, right side of neck, left ala nasi and left temporal and zygomatic areas (Fig. 2). The patient stated that these nonlinear lesions had developed after puberty. She could not recall similar skin changes in her family members, but she no longer had contact with them.

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Histological examination of biopsies obtained from a segmental lesion in the right eyebrow area and from one of the disseminated papules localized on the right cheek showed a hyperpigmented epidermis with acanthosis and papillomatosis. A pronounced fibrous stroma was noted in the dermal component, thus representing typical features of DPN, although these did distinguish DPN from SK.

A literature review did not reveal any other case of DPN showing a segmental arrangement. In our patient, most of the facial lesions were distributed in a

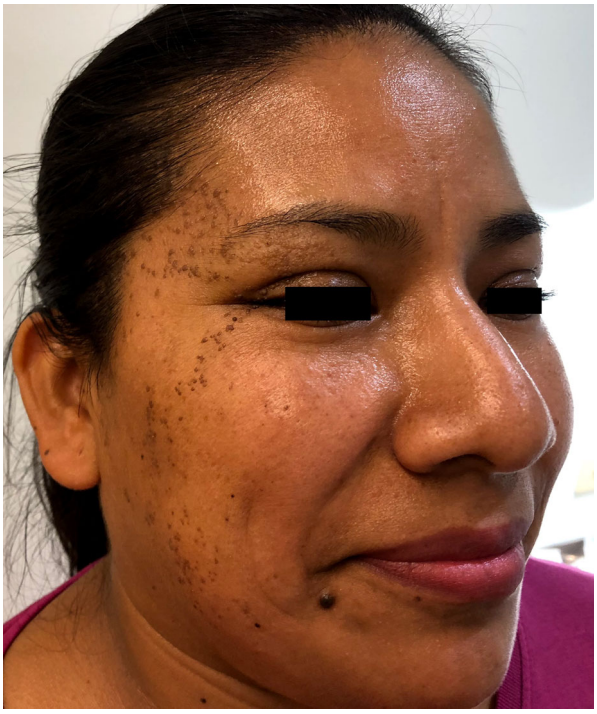

Figure 1 Linear lesions of dermatosis papulosa nigra involving exclusively the right side of the face, with additional non-segmental papules on the ear and neck and below the lower lip.

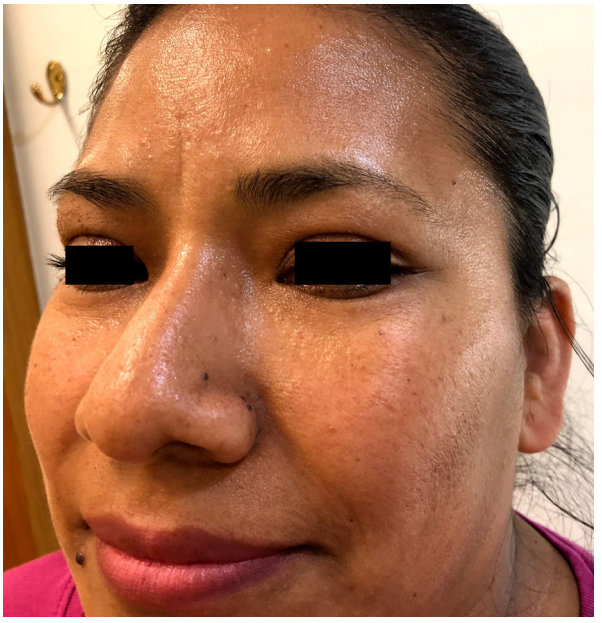

Figure 2 Nonsegmental papules of similar appearance around the left ala nasi and in the left temporal and zygomatic areas.

linear pattern, which may be taken as an example of superimposed segmental mosaicism. Common skin disorders with a polygenic background sometimes show a linear or otherwise segmental arrangement of rather pronounced involvement that may either represent an isolated disorder or be superimposed on less severe, disseminated, symmetrically distributed lesions of the same disorder.[5] This phenomenon has tentatively been explained by the '$n + 1$' rule.[6] For example, a patient may have both unilateral linear psoriasis and common, nonsegmental plaques of the same disorder. The unknown number of inherited predisposing heterozygous alleles would be n, whereas those within the superimposed segmental areas would be $n + 1$. Either postzygotic loss of the corresponding wild-type allele at one of the inherited predisposing genes may have happened at an early developmental stage, or a postzygotic new mutation may have occurred at an additional predisposing gene locus.[6] Reports indicating superimposed segmental mosaicism have been published for many common cutaneous diseases, including pustular psoriasis, atopic dermatitis, lichen planus, dermatomyositis, systemic lupus erythematosus, granuloma annulare, erythema multiforme, graft-versus-host disease, acne vulgaris and vitiligo.[5]

Future molecular research on superimposed segmental manifestation of polygenic skin diseases may show whether the hypothetical $n + 1$ rule holds true, and whether DPN can be added as a further example to this group of common disorders.

References

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CPD questions

Learning objective

To gain up-to-date knowledge of the aetiology and histological features of dermatosis papulosa nigra.

Question 1

Which of the following statements correctly describes dermatosis papulosa nigra?

(a) Congenital condition manifesting in childhood.
(b) Acquired condition manifesting in adulthood.
(c) Acquired condition manifesting in childhood.
(d) Congenital condition manifesting in adulthood.
(e) Dermatosis related to sun exposure.

Question 2

Which of the following conditions may dermatosis papulosa nigra resemble histologically?

(a) Common wart.
(b) Seborrhoeic keratosis.
(c) Basal cell carcinoma.
(d) Dermatofibroma.
(e) None of the above.

Instructions for answering questions

This learning activity is freely available online at http://www.wileyhealthlearning.com/ced

Users are encouraged to

- Read the article in print or online, paying particular attention to the learning points and any author conflict of interest disclosures
- Reflect on the article
- Register or login online at http://www.wileyhealthlearning.com/ced and answer the CPD questions
- Complete the required evaluation component of the activity

Once the test is passed, you will receive a certificate and the learning activity can be added to your RCP CPD diary as a self-certified entry.

This activity will be available for CPD credit for 2 years following its publication date. At that time, it will be reviewed and potentially updated and extended for an additional period.