Linear Arrangement of Multiple Deep Penetrating Nevi

Report of First Case and Review of Literature

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Background: Deep penetrating nevus is a recently described variant of melanocytic nevi with clinical and histopathological features that may be confused with malignant melanoma, blue nevus, pigmented Spitz nevus, or congenital melanocytic nevus. We report a case with linear arrangement of multiple deep penetrating nevi. To our knowledge, such presentation has never been reported in the literature.

Observations: We describe a patient with multiple darkly pigmented lesions in the right periauricular area, above and behind the ear. The histopathological features of these lesions were consistent with deep penetrating nevus.

Conclusions: To our knowledge, this is the first report of linear arrangement of multiple deep penetrating nevi. We consider this case a unique presentation of deep penetrating nevus.

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DEEP PENETRATING nevus (DPN) is a recently described pigmented melanocytic nevus. Clinically, DPN is a solitary darkly pigmented lesion, usually less than 1 cm in diameter, most commonly found on the face, neck, or shoulder. Deep penetrating nevus must be recognized because it possesses histopathological features that may be alarming and can be mistaken for malignant melanoma. It should also be differentiated from other pigmented nevi, namely, blue nevus, pigmented Spitz nevus, or congenital melanocytic nevus. We report a case of multiple deep penetrating nevi showing a linear distribution. To our knowledge, this is the first reported case with such presentation.

On examination, 7 slightly elevated, smooth-surfaced, well-defined, darkly pigmented lesions were found in a linear distribution in the periauricular area above and behind the right ear in the area supplied by the lesser occipital nerve. The diameter of the lesions ranged from 2 to 8 mm. One of the lesions was surrounded by 3 tiny (diameter, <1 mm) satellite lesions (Figure 1). All lesions were excised and histopathologically examined. All lesions showed similar histopathological features and were symmetrical, well-demarcated, and wedged-shaped with the apex in the subcutaneous fat. The lesions consisted of melanocytes with oval monomorphous nuclei and abundant pale cytoplasm containing dusty particles of melanin. Although most of the melanocytes had monomorphous nuclei, some had large pleomorphic nuclei, as seen in the Spitz nevus. Low-grade cytological atypia consisting of variation in nuclear size, shape, and hyperchromasia were observed. Mitotic figures were not present. The melanocytes were mainly arranged in nests and fascicles throughout the lesions. The melanocytes were present between the collagen bundles and around the hair follicles, sweat glands, blood vessels, and nerves. Some nests of melanocytes were also seen at the dermoeidermal junc-

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tion. Numerous pigment-laden macrophages were evident, especially in the deeper part of the lesions. In addition, perivascular lymphocytic infiltrate was present. No fibrosis was observed. The histopathological features were consistent with compound DPN (Figure 2). Histopathological examination findings of the tiny satellite lesions showed a few nests of melanocytes with oval monomorphous nuclei in the upper dermis. Some of these nests were present around the blood vessels.

Physical examination of the rest of the patient’s body skin showed only the presence of a few normal-appearing acquired nevi.

In 1989, Seab et al described a series of patients with a pigmented melanocytic lesion that they termed deep penetrating nevus. This was followed by other series emphasizing the clinical and histopathological features of this nevus and stressing the need to differentiate it from malignant melanoma. Clinically, DPN is an elevated, dome-shaped, well-demarcated, deeply pigmented lesion less than 1 cm in diameter. The most common locations of DPN are the face, neck, and shoulders of patients between the ages of 10 and 30 years. We describe a patient with linear melanocytic lesions in which the histopathological findings were consistent with DPN. Clinically, our patient had 7 slightly elevated, smooth-surfaced, deeply pigmented lesions in a linear distribution in the right periauricular area above and behind the ear. One of the lesions was surrounded by 3 tiny satellite lesions.

To our knowledge, such linear arrangement of multiple deep penetrating nevi has never been reported. Furthermore, the presence of satellite lesions with DPN has not been described. We consider our case to be unique in its presentation.

A review of the literature revealed descriptions of other melanocytic lesions presenting with a linear or otherwise segmental distribution. The histopathological features of DPN are alarming, and Seab et al reported that 29% of patients were falsely diagnosed as having melanoma. However, melanoma lesions are poorly circumscribed and not symmetrical. Moreover, the melanocytes in melanoma are present in the epidermis above the dermoepidermal junction. Along the dermoepidermal junction, there is a predominance of melanocytes arranged as solitary units over melanocytes in nests. Moreover, there is asymmetry of the melanin distribution. In addition, mitosis is usually present in malignant melanoma.

The distinction between DPN and cellular blue nevus and pigmented congenital melanocytic nevus has been made in previous studies. The histopathological features differentiating these nevi from DPN are summarized herein. In cellular blue nevi, there are no nests of melanocytes at the dermoepidermal junction. In addition, dendritic melanocytes, the characteristic cells of blue nevi, are not evident in DPN. Congenital melanocytic nevi are present since birth, and...
there are no nuclear atypia of the melanocytic nevus cells, but there is maturation of the nuclei with deeper descent into the dermis. Furthermore, melanocytes with large pleomorphic nuclei resembling those seen in Spitz nevus are present in DPN and absent from congenital melanocytic nevus and cellular blue nevus.

The presence of satellite lesions has been described in association with other melanocytic nevi, including blue nevus\textsuperscript{10} and congenital nevus\textsuperscript{17}, however, DPN associated with satellite lesions as seen in our patient has not been described. The histopathological examination of the satellite lesions revealed dermal nests of nevus cells arranged around the blood vessels.

The cause of the unique distribution of DPN in our reported case is unclear; however, as suggested in other melanocytic nevi,\textsuperscript{8,7} one may postulate that a genetic defect determines the appearance of DPN. The linear distribution is a clinical manifestation of mosaicism.\textsuperscript{18} Accordingly, we hypothesize that a postzygotic somatic mutation occurring early in embryogenesis may give rise to a mutant clone carrying a major gene that is responsible for the development of DPN. Later in life, the pre-determined development of DPN is triggered by an unknown stimulus, perhaps UV radiation. Interestingly, a concept of dichotomous types of segmental manifestations in autosomal dominant skin disorders has recently been proposed by Happle.\textsuperscript{19} Type 1 reflects heterozygosity for the underlying postzygotic new mutation and shows a degree of severity corresponding to that encountered in the nonsegmental phenotype. Type 2 results from an early postzygotic loss of heterozygosity of the corresponding wild-type allele, resulting in a population of cells that are homozygous or hemizygous for the underlying gene. As a characteristic feature, the segmental type 2 is superimposed on a milder, nonsegmental, heterozygous manifestation of the same trait. Although the mode of transmission of DPN is speculative at this time, the unique distribution in our case is more consistent with type 1 segmental involvement rather than the concept of loss of heterozygosity, because the remaining skin appeared to be healthy and the family history was negative.

Future molecular studies concerning DPN will be needed to confirm any of these concepts and to help in determining the origin of this nevus.

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\textbf{REFERENCES}


