The deep penetrating nevus

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The deep penetrating nevus (DPN), also known as the plexiform spindle cell nevus, is a pigmented lesion that commonly arises on the head and neck in the first few decades of life. Histopathologically, the DPN is wedge-shaped and contains melanocytes that exhibit deep infiltration into the dermis. Given these features, DPN may clinically and histopathologically mimic malignant melanoma, sparking confusion about the appropriate evaluation and management of these lesions. The goal of this review is to summarize the clinical and histopathological features of DPN and to discuss diagnostic and treatment strategies for dermatologists. (J Am Acad Dermatol 2014;71:1234-40.)

Key words: deep penetrating nevus; dermatopathology; malignant melanoma; melanocytic tumor of uncertain malignant potential; nevus with focal atypical epithelioid components; plexiform spindle cell nevus.

The deep penetrating nevus (DPN) is a darkly pigmented melanocytic lesion that exhibits deep infiltration into the reticular dermis.[1] It has gained recognition over the past couple of decades because its histologic appearance is commonly confused with malignant melanoma. This nevus does not have a consistent morphology and because of its often sudden appearance and dark color, its appearance may raise concern for a malignant process among both practitioners and patients. Although the pathology literature is well-versed in the histologic features of DPN, the dermatology literature lacks a comprehensive clinical summary, and dermatologists are often forced to make clinical decisions based on isolated case reports and small series.

The goal of this review is to summarize the clinical and histologic features of DPN and to discuss diagnostic and treatment issues for these elusive lesions.

BACKGROUND

The DPN was first described in 1989 by Seab et al,[1] who reported 70 patients with a darkly pigmented nevus composed of loosely organized nests of melanocytes with deep extension into the reticular dermis. In 1991, Barnhill et al[2] described a lesion with similar characteristics, which they named the "plexiform spindle cell nevus." It was later accepted that these 2 reports were describing the same melanocytic lesion, and the naming of the nevus by Seab et al[1] was accepted. Since then, nearly 400 lesions have been reported in the English-language literature.

CLINICAL PRESENTATION

Although the age at presentation is broad (0-77 years) (Table I), DPN tend to occur in a younger patient population, most frequently in the first 3 decades of life.[1,3,4] Congenital onset has been reported,[3,5-7] and lesions rarely arise in patients older than 50 years.[8] Females appear to have a slightly higher incidence (57.4%) compared with males (42.4%).[1-3,5,6] DPN occur on the head and neck in approximately 35% of cases, on the trunk in 25%, and on the upper extremities in 20.5%. Less commonly, DPN occur on the lower extremities, and they have only rarely been described on distal extremities[3,4] or mucosal surfaces.[9,10]

The majority of DPN are darkly pigmented dome-shaped papules or nodules that are less than 1 cm in diameter.[8,11] Typically, DPN present as a solitary lesion, however, a linear arrangement of multiple DPN over the preauricular cheek has

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been reported.[12] The lesions are most frequently black, blue, dark brown, or gray (or often a combination of these colors) (Fig 1) but have been rarely noted to be red or pink.[3] Patients may report a new or changing lesion because they are almost always acquired. Most DPN are asymptomatic, although pain or dysesthesia have rarely been reported.[13] The vast majority of patients have no personal or family history of melanoma. A DPN developing within a medium-sized congenital nevus has been reported in a 3-month old infant.[14]

CAPSULE SUMMARY

- The deep penetrating nevus is a melanocytic lesion that may clinically and histopathologically mimic malignant melanoma.
- This review summarizes the clinical and histopathological features and the outcomes of deep penetrating nevus that have been published in the literature.
- The deep penetrating nevus is a controversial neoplasm that has rarely been associated with recurrence or lymph node involvement. More research is needed to explore the behavior of these lesions.

DIAGNOSIS

DPN should be considered in any young patient presenting with a new or changing darkly pigmented papule or nodule. Clinically, these lesions appear melanocytic in nature and only rarely have been confused with seborrheic keratoses or hemangiomas.[3] The differential diagnosis for such a melanocytic lesion is broad and includes blue nevus, Spitz nevus, pigmented spindle cell nevus, atypical nevus, and malignant melanoma (Table II).

The dermoscopic features of DPN have not been well established. One patient was noted to have a symmetric lesion with a homogenous "negative" globular pattern at baseline.[15] As the lesion rapidly expanded, the globular pattern became confluent and cobblestone-like structures were noted centrally, prompting a biopsy specimen that confirmed the diagnosis.[15] Subsequently, the dermoscopic features of 2 DPN were described as "polychromatic," a term used when 3 or more colors are present.[16] Given these nonspecific clinical and dermoscopic findings, establishing the diagnosis of a DPN purely based on appearance with or without a dermoscope is nearly impossible, and pathologic examination of the lesion is essential.

An excisional biopsy specimen is the ideal way to evaluate DPN to examine the lesion in its entirety. Heavy pigmentation may be noted on gross examination of the excised tissue, a finding that is also not unusual to see with blue nevi.[3] If a "scoop shave" removal is performed, the clinician is occasionally alarmed to see dark pigment at the deep edge of the biopsy site, which might raise concern for malignant melanoma. Therefore if the clinical suspicion for DPN is high, the best method of diagnosis is via a narrow excision.

HISTOLOGIC FEATURES

The DPN is a sharply demarcated lesion, and at low power is symmetric and wedged-shaped with a broad base that runs parallel with the epidermis and an apex that is oriented towards the subcutaneous adipose.[1-3,5,6,17] It consists of often vertically oriented, loose fascicles and nests of spindle-shaped and epithelioid melanocytes extending into the reticular dermis in a plexiform pattern (Figs 2 and 3). Melanophages can be found in focal interstitial areas among the groups of nests and fascicles.[1-3,5,6,17] There are typically a few nests or melanocytes in a lentiginous pattern at the dermoepidermal junction.[1,3] The papillary dermis is frequently spared[8]; however, these lesions are often combined with a common dermal or congenital nevus in the epidermis and papillary dermis. Melanocytes tend to spread along hair follicles, blood vessels, sweat glands, and nerves, and have been found to infiltrate the arrector pili muscles in up to 88% of cases.[3] Subcutaneous involvement is common. Mature lymphocytes are responsible for a mild to moderate inflammatory reaction in approximately 75% of cases.[6]

Melanocytes in DPN have abundant pale cytoplasm, small nucleoli, smudged chromatin, and large nuclear pseudoinclusions.[1-3,5,6,17] Occasional low-grade cytologic atypia and random nuclear pleomorphism are commonly found, often seen in the deepest part of the lesion.[1-3,5,6,17] Although mitoses are rare, 0 to 1.2 mitoses/mm^2 is possible.[5]

Approximately two thirds of DPN have a combined appearance[3]; most often acquired nevi are associated.[3] Frequently, the acquired nevus can be seen occupying the most superficial part of the lesion, whereas the characteristics of the DPN can be seen extending into the reticular dermis. Less commonly, components of blue nevi and Spitz nevi can be found in association with DPN.[5] Some experts believe that the term "deep penetrating nevus" should be reserved for those lesions that are not combined.[18]