Answer to Dermacase *continued from* **page 1361**

5. Speckled lentiginous nevus

A speckled lentiginous nevus (SLN), or *nevus spilus* (from the Greek word *spilos*, meaning *spot*), clinically appears as a hyperpigmented macule or patch containing scattered, dark, macular or papular foci. Speckled lentiginous nevi are identified in less than 0.2% of neonates,[1] 1% to 2% of white schoolchildren,[2] and 2% to 3% of white adults,[3,4] although no data are available on the prevalence in individuals with darker skin. There is no sex predilection, although there is a distribution predilection for the trunk and lower limbs.[4]

Speckled lentiginous nevi can be congenital or acquired lesions, and the distinction is based on patient history and histology. A congenital SLN is present at birth or soon after, might have segmental or zosteriform patterns of distribution reflecting embryonic development, and shows histologic features consistent with a congenital skin lesion.[5] Speckled lentiginous nevi can be overlooked at birth owing to their light coloration, and might become more obvious and deeply pigmented months to years later, making it difficult to distinguish congenital from acquired lesions based on history alone. In the case described above, the SLN was indeed present since birth and showed histologic features consistent with a congenital skin lesion.

Diagnosis

Clinical appearance and patient history typically provide sufficient information to make the diagnosis of SLN. Clinically, an SLN appears as an evenly hyperpigmented macule or patch containing dark, scattered foci. The background macule or patch can range in size from less than 1 cm to greater than 10 cm.[6] A Wood lamp might be needed to visualize the background patch if it is weakly pigmented. The differential diagnosis of SLN includes nevomelanocytic nevus, lentigo, café au lait spot, and malignant melanoma.

Human skin is made up of several types of cells, including melanocytes, which contain melanin pigment and account for the colour of skin. In normal skin, single melanocytes are mainly found interspersed among other cells along the epidermal basement membrane, and less commonly within the dermis and upper epidermis. A nevomelanocytic nevus, also known as a common mole, appears clinically as a small, round, uniform, pink to dark-brown macule, papule, or plaque. Histologically, melanocytes are in small, circumscribed, organized nests within the epidermis (junctional nevomelanocytic nevus), dermis (intradermal nevomelanocytic nevus), or both (compound nevomelanocytic nevus). They typically appear in childhood or young adulthood and are more frequent in the second and third decades of life, appearing commonly on sun-exposed areas of skin.[7]

Lentigines are formed by an increased number of neighbouring melanocytes at the epidermal basement membrane that do not form nests. They can be congenital or acquired lesions that typically appear as sharply circumscribed, light-brown or dark-brown macules, otherwise known as *freckles*. Macules can be isolated or aggregated (present as a cluster of macules), and can be found on the skin, nails, and mucus membranes. Aggregated lentigines can mimic the appearance of an SLN. A Wood lamp can be useful in determining the existence of a background hyperpigmented patch, which is typical of an SLN but not of aggregated lentigines.[8]

Café au lait spots are formed by a normal number of melanocytes in the skin that contain increased amounts of melanin pigment. Clinically, they are flat, sharply circumscribed, and evenly coloured light-brown macules or patches without the dark foci that are typically seen in SLN. They are frequently present at birth, become more numerous as the child grows, and tend to grow in proportion to the overall growth of the child. The presence of numerous café au lait spots can be a marker for certain syndromes such as neurofibromatosis type 1.[9]

Malignant melanoma describes the aberrant proliferation of single melanocytes from within a nevus or from solitary cells within the skin. Several clinical and histologic subtypes of melanoma exist, although they are rare in childhood and adolescence. It can be clinically difficult to distinguish a benign nevomelanocytic nevus from an early melanoma. Clinical suspicion of melanoma is raised with a new lesion, a change in size of an existing lesion, the presence of nonuniform features such as a variation or change in colour, asymmetry, or border irregularity.[10]

Evolution

An SLN can evolve over the lifetime of the individual. Specifically, new hyperpigmented elements within the background macule or patch might appear, or existing elements might evolve over time. If an element appears nonuniform, appears unusual compared with the others, or changes over time, the development of an early melanoma should be ruled out.

The absolute risk of developing a malignant melanoma from an SLN is poorly defined. In an early study, Kopf et al identified nevi spili in 4.8% of a cohort of 105 white adults with melanoma compared with 2.3% of a cohort of 601 melanoma-free adults; however, this difference was not statistically significant.[11] Nevertheless, numerous case reports have described the development of malignant melanoma from SLN.[12-19] A systematic review including 14 studies and 6571 patients with congenital melanocytic nevi reported that the overall risk of developing malignant melanoma was 0.7%.[20] Further, the risk of developing melanoma and the rate of a fatal course increased for larger congenital melanocytic

nevi (>40 cm in diameter).[20] Other smaller studies have reported an incidence of melanoma of about 1% to 2%.[21,22] Larger studies with long-term follow-up are necessary to calculate the absolute risk of melanoma development from SLN, although it is likely small.

Management

Patients with speckled lentiginous nevi should be informed that, as with most melanocytic lesions, there is a small lifetime risk of malignancy. For this reason, their lesions should be assessed by physicians annually.[17] Dermoscopy is a noninvasive method of screening for atypical changes in the hyperpigmented foci within the background macule or patch of the SLN. Photographic documentation can be used to monitor changes in the SLN over time. Biopsy should be reserved for lesions in which there is suspicion of malignancy, such as those with atypical elements including changes in size and colour, border irregularities, or growth of an element within the SLN.[23] When in doubt, referral to a dermatologist is warranted.

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Competing interests
None declared

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