Speckled Lentiginous Nevus
Within the Spectrum of Congenital Melanocytic Nevi

Julie V. Schaffer, MD; Seth J. Orlow, MD, PhD; Rossitza Lazova, MD; Jean L. Bolognia, MD

Background: Currently, there is disagreement as to whether speckled lentiginous nevi (nevi spili) are congenital or acquired pigmented lesions. Part of this controversy is related to the natural history of these lesions that often present at birth as hyperpigmented patches and then take several years to reach their more readily recognized spotted form. Arguments in favor of speckled lentiginous nevi as a subtype of congenital nevi include the following observations: multiple reports of lesions present at birth or noted soon thereafter; patterns of distribution reflecting embryonic development; hamartomatous behavior with various types of nevi (eg, junctional nevi, blue nevi, and Spitz nevi) presenting in the same lesion over time; and histologic features of congenital melanocytic nevi within the spots. Herein we present additional evidence for the congenital nature of speckled lentiginous nevi.

Observations: Ten patients are described with congenital pigmented lesions that had the clinical appearance of speckled lentiginous nevi in whole or in part. These lesions either evolved and acquired an appearance more suggestive of “classic” congenital nevi, or they existed as “hybrid” lesions with portions appearing as classic congenital nevi adjacent to or admixed with portions appearing as speckled lentiginous nevi. On histologic examination, biopsy specimens from the spots within these lesions showed features of congenital melanocytic nevi.

Conclusions: These 10 cases, along with the arguments outlined above, provide strong support for the hypothesis that speckled lentiginous nevi are a subtype of congenital melanocytic nevi.

Arch Dermatol. 2001;137:172-178

In clinical studies involving children and adults, speckled lentiginous nevi (nevi spili) were shown to have a prevalence in the general population similar to that of congenital melanocytic nevi.1 For example, in 1 series of primarily adult patients (n=601), 2.3% had a speckled lentiginous nevus larger than 1.5 cm in diameter,2 and in 3 large series of school-aged children and adolescents (n=3654), prevalence rates varied from 1.3% to 2.1%.3-5 However, much lower prevalence rates have been reported in series of newborns. Rhodes and Mihm4 noted that only 2 of 1118 newborns had a clinically evident speckled lentiginous nevus. Based on these findings as well as the findings from 2 other series of 4641 and 10588 newborns in which no speckled lentiginous nevi were observed, Rhodes and Mihm5 stated that speckled lentiginous nevi are acquired lesions. In contrast, of 26 patients described in 4 recent series,6-12 21 (81%) had lesions that were present at birth or noted during early infancy; these series included small lesions, with at least 40% measuring less than 6 cm in diameter.

One explanation for the controversy as to whether speckled lentiginous nevi are congenital or acquired is based on their natural history. These lesions can appear at birth as lightly colored café-au-lait macules and may be subtle in their clinical presentation, whereas the more obvious pigmented macules and papules develop over a period of months to years to decades.6,11,13-15 Furthermore, it is clear that some classic congenital nevi only become apparent after several months to years; for example, Mackie et al15 defined a congenital nevus, also referred to as an early onset nevus, as arising within the first 2 years of life. The term congenital can thus encompass lesions that are programmed from birth as well as those overtly apparent at birth.

We contend that speckled lentiginous nevi...
are congenital lesions that may take several years to develop their multiple spots.

There are several additional arguments in favor of speckled lentiginous nevi representing a subset of congenital nevi (Table 1). Occasionally, speckled lentiginous nevi involve extensive areas of the body, and when this occurs a sharp demarcation at the midline may be seen. The overall shape may be blocklike or linear, and the distribution patterns are often referred to as zosteriform. In addition, the linear lesions may follow the lines of Blaschko, a pattern known to originate during embryonic development. A divided form of speckled lentiginous nevus that involves both the upper and lower eyelids also has been described. This phenomenon reflects the probable timing of nevus formation during embryonic development, occurring after eyelid fusion in the eighth to ninth week of gestation but before eyelid reopening in the sixth month of gestation. More classic congenital divided melanocytic nevi of the eyelid have also been reported.

The macular and papular spots of a speckled lentiginous nevus often range from 1 to 3 mm in diameter but occasionally are larger. The speckles can have a wide spectrum of clinical appearances as well as histologic features. They range from lentigines to junctional, compound, and intradermal nevi to Spitz and neural nevi. It is important to note that different types of spots may appear at different times during the life of a speckled lentiginous nevus. For example, at age 3 years, 1 child was described as having multiple Spitz nevi within his speckled lentiginous nevus, but when examined at age 13 years, he had multiple blue and compound nevi within the lesion (S.J.O., personal observation, 1993). In some reports, progressive changes have been related to factors such as sun exposure and pregnancy. In other words, speckled lentiginous nevi exhibit hamartomatous behavior, as do classic congenital nevi where surface irregularities, neurotization, and/or superimposed papules and nodules develop over time.

Classically, the background hyperpigmentation of a speckled lentiginous nevus is described histologically as having the features of a lentigo or cafe au lait macule. However, there are reports of nevus cells being present in biopsy specimens of this background hyperpigmentation; this argues against using the histologic features of the background hyperpigmentation as the sole criterion for the diagnosis of this particular pigmented lesion. In addition, the spots of a speckled lentiginous nevus are classically described as superimposed junctional or compound nevi. Histologic features associated with congenital nevi were described in reports of biopsy specimens of the speckles, ie, collections of nevus cells in the reticular dermis and within adnexal structures. Additional investigations of the pathologic features of the spots within speckled lentiginous nevi have confirmed this finding.

We describe 10 patients with congenital pigmented lesions that had the clinical appearance of a speckled lentiginous nevus in whole or in part...
The lesions either evolved into a more classic congenital nevus, or existed as hybrid lesions with a speckled lentiginous nevus in contiguity with a classic congenital nevus. These cases therefore add 2 more arguments in support of our assertion that speckled lentiginous nevi represent a subtype of congenital melanocytic nevus (Table 1).

**REPORT OF CASES**

**CASE 1**

A white male infant was initially seen at age 6 months for evaluation of a pigmented lesion on the right upper abdomen (Figure 1A). A patch of hyperpigmentation had been noted during his first month of life, followed by the appearance of at least 10 smaller brown macules of varying sizes within this area. The clinical appearance of the pigmented lesion at age 6 months was that of a speckled lentiginous nevus (nevus spilus), and a biopsy specimen was obtained of one of the slightly elevated brown papules. By age 12 months, the lesion had darkened slightly throughout, but its increase in size was solely in proportion to the growth of the child (Figure 1B). Thirteen months later, at age 2 years, the nevus had the clinical appearance of a more classic congenital melanocytic nevus (Figure 1C). It was now medium brown and elevated in its entirety. The border was slightly jagged, and there was evidence of perifollicular hypopigmentation. Some of the original spots were still visible, whereas others had blended into the background color.

The child was last examined at age 9 years. The nevus then measured 6.5 × 2.8 cm and had an appear-
ance similar to that seen at age 2 years. The lesion was completely excised shortly after this evaluation. The patient’s only medical problem was an attention deficit disorder. There was no family history of congenital melanocytic nevi or melanoma.

Histologic examination of the biopsy specimen of one of the darker papules obtained at age 6 months (Figure 1A) showed nests of uniform melanocytes in the superficial dermis and at the dermoepidermal junction. Focally, a few nests and cords of melanocytes were seen around adnexal structures in the upper reticular dermis. The diagnosis was compound melanocytic nevus with congenital features. The surgical specimen from the excision at age 9 years showed nests of small and monomorphic melanocytes at the dermoepidermal junction and within the reticular dermis, with many melanocytes concentrated around blood vessels and adnexal structures; the diagnosis was compound congenital melanocytic nevus (Figure 2).

CASE 2

A 7-year-old white girl was seen for evaluation of a congenital nevus on her left hand. The patient’s parents reported that they had not noted any specific changes in the lesion in several years. The patient’s only medical problem was mild asthma. There was no family history of congenital melanocytic nevi, but there was a possible history of cutaneous melanoma in a paternal uncle.

On physical examination, a 3 × 4-cm congenital melanocytic nevus involving the left second, third, and fourth fingers as well as the skin overlying the third and fourth metacarpophalangeal joints was observed. Proximally on the third finger was the darkest portion of the nevus; it measured 1 × 1.5 cm (Figure 3) and was the only area of the nevus that was obviously elevated or contained an increased number of terminal hairs. This portion of the nevus was removed by staged excision between the ages of 7 and 9 years because of cosmetic concerns. More peripherally, there was macular hyperpigmentation of the skin with superimposed darker brown macules of various sizes (Figure 3).

Histologic examination of the surgical specimen from the third finger showed nests of monomorphic melanocytes at the dermoepidermal junction and within the dermis, with some melanocytes concentrated around blood vessels and adnexal structures; the diagnosis was compound congenital melanocytic nevus. The surgical specimen from the fourth finger showed nests of melanocytes at the dermoepidermal junction. Focally, small and uniform melanocytes were seen around blood vessels of the superficial plexus. The diagnosis was compound melanocytic nevus with congenital features.

For this particular pigmented lesion, there is disagreement regarding nomenclature (speckled lentiginous nevus vs nevus spilus) as well as age of onset (congenital vs acquired). The terminology controversy dates back to the use of the term nevus spilus in the late 19th and early 20th centuries to describe a uniformly pigmented brown patch. Several writings from this period are often cited to support the argument that use of the term nevus spilus creates confusion.

Although these early references contain somewhat nonspecific descriptions such as (1) a birthmark with a smooth, soft surface, (2) a lesion whose clinical description can be completely confused with that of a lentigo, and (3) a “permanent alteration of the color of the skin, either of its whole surface or of some of it only, without elevation or desquamation,” it is clear that there was never any mention of superimposed, more darkly pigmented macules or papules.

In 1952, Ito and Hamada reintroduced the term nevus spilus to describe a hyperpigmented patch with superimposed darker speckles, and its use was subsequently reinforced by Cohen et al. Then, in the mid-1970s, the term speckled lentiginous nevus first appeared as a name for pigmented lesions with a background of macular hyperpigmentation containing smaller, darker macules and/or papules. In a review of MEDLINE citations over the past 10 years (1990-2000), the 2 terms have been used in a ratio of approximately 1.5 nevus spilus to 1 speckled lentiginous nevus.

We favor the term speckled lentiginous nevus not simply from a historical perspective or because translation of the Greek root spilos is “spot,” not “spotty” or “spotted,” but as a means of emphasizing the biological behavior of this pigmented lesion. A speckled lentiginous nevus can be likened to a melanocytic garden, and within this garden a variety of lesions can grow, from junctional nevi to blue nevi to melanoma. Unfortunately, as recently as 10 to 15 years ago, a nevus spilus was viewed (incorrectly) as a lesion with no malignant potential, and even today the histologic features of the spots of a nevus spilus are usually described simply as those of a junctional nevus. As previous authors have pointed out, use of the term speckled lentiginous nevus serves to broaden both the clinical and histologic spectrum. Finally, we believe that the practice of referring to a smaller lesion as a nevus spilus and a larger lesion as a speckled lentiginous nevus simply adds to the confusion.

Recent reports of cutaneous melanomas developing within speckled lentiginous nevi have increased the awareness of these lesions as potential precursors of melanoma. There have been at least 20 such cases reported, a few of which have been fatal. The background lesions varied in size from 2 cm in diameter to extensive zosteriform lesions, and some had speckles with atypical cytologic features. What remains to be determined is whether the surface area, the number or type of superimposed nevi, or the presence of atypical cytologic features correlates with the risk of developing melanoma. Recently, the level of concern regarding these lesions has become similar to that assigned to classic congenital nevi of similar size. From suggested that speckled lentiginous nevi may actually present a greater risk, noting that
of more than 2000 melanomas seen at her pigmented lesion clinic during a 15-year period, more arose from speckled lentiginous nevi (n = 3) than from large classic congenital melanocytic nevi (n = 1).

The cases we report here represent 2 different examples of the connection between speckled lentiginous nevi and congenital melanocytic nevi. In the first example, the hybrid pigmented lesions in patients 2 through 10 had portions with the clinical and histologic features of a classic congenital melanocytic nevus either adjacent to (Figure 3 and Figure 4) or admixed with (Figure 5) portions that had the appearance of a speckled lentiginous nevus (of note, there are at least 2 previously described cases of the adjacent type of hybrid lesion12,25). In the second example (Figure 1), a typical speckled lentiginous nevus evolved into a more classic congenital melanocytic nevus with a few spots, a lesion that might be referred to by some clinicians as a spotted grouped nevus. More specifically, the term spotted grouped pigmented nevus has been used to describe a variant of congenital melanocytic nevus that has speckles in a perifollicular and/or perieccrine distribution.74-76 It is our opinion that both speckled lentiginous nevi and spotted grouped pigmented nevi fall within the spectrum of congenital melanocytic nevi.

If one accepts the premise that speckled lentiginous nevi are a subtype of congenital melanocytic nevi, then how should such lesions be managed? Our recommendation is to perform baseline photographic documentation followed by serial clinical observation and education of the patient and family regarding the clinical signs of melanoma. In general, speckled lentiginous nevi are much easier to follow clinically than are classic congenital nevi, which are often uniformly elevated, dark brown or black, and covered by dense terminal hairs. Although we do not advocate excision of speckled lentiginous nevi (which would require removal of the entire hyperpigmented patch to eliminate the field defect), we do recommend that a biopsy of any suspicious area be performed.

We have presented 10 cases with clinical histories as well as histologic features that support the hypothesis that speckled lentiginous nevi are a subtype of congenital melanocytic nevi. These patients had lesions that existed in either spatial or temporal contiguity with classic congenital nevi. These observations along with those outlined in Table 1 provide a strong argument in support of this contention. With regard to terminology, we prefer the use of speckled lentiginous nevus with a qualifier, such as Spitz nevi within a speckled lentiginous nevus or blue nev within a speckled lentiginous nevus.

<table>
<thead>
<tr>
<th>Patient No./Age/Sex</th>
<th>Age at Onset</th>
<th>Location</th>
<th>Clinical History</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/9 y/M 1/6 mo/F</td>
<td>First month of life</td>
<td>Right upper abdomen</td>
<td>See text (Figure 1).</td>
</tr>
<tr>
<td>2/7 y/F 3/19 mo/F</td>
<td>Birth</td>
<td>Dorsum of left hand</td>
<td>A portion appeared as an area of hyperpigmentation and hypochromia; an adjacent less pigmented portion had multiple superimposed darker macules. Over time the portion in the inguinal crease darkened, and an increase in size was proportional to the growth of the child (Figure 4).</td>
</tr>
<tr>
<td>4/14 mo/F 6/6 mo/M</td>
<td>Birth</td>
<td>Flank</td>
<td>Initially appeared as a patch of tan to light brown hyperpigmentation with 3 superimposed darker macules; during the first year of life, multiple darker brown papules developed within the lesion.</td>
</tr>
<tr>
<td>5/3 mo/M 7/1 mo/M</td>
<td>Birth</td>
<td>Upper back</td>
<td>Appeared as an area of hyperpigmentation with superimposed darker brown to black macules, papules, and plaques; with some nevus-like components and hypertrichosis within the larger plaques. Multiple satellite lesions were present.</td>
</tr>
<tr>
<td>6/6 y/M 7/1 mo/M</td>
<td>Birth</td>
<td>Lower back and buttocks</td>
<td>Appeared as an area of café au lait-like tan hyperpigmentation with numerous (&gt;100) superimposed macular or slightly elevated lesions ranging from 1 mm to 5 cm and varying from tan to dark brown, with hypochromia within the larger, darker plaques. An increase in size was proportional to the growth of the child.</td>
</tr>
<tr>
<td>8/6 mo/M 9/2 mo/M</td>
<td>Birth</td>
<td>Lower back, buttocks, and genitals</td>
<td>Initially appeared as an area of light brown macular hyperpigmentation with multiple superimposed papules, nodules, and plaques varying from red to black. Over time the entire lesion became elevated, and an increase in size was proportional to the growth of the child.</td>
</tr>
<tr>
<td>10/4 mo/M 10/1 y/F</td>
<td>Birth</td>
<td>Lower back and buttocks</td>
<td>Initially appeared as a patch of medium brown hyperpigmentation with superimposed scattered dark brown to black papules and plaques; at age 3 months, several 1- to 2-cm firm nodules rapidly developed within the lesion.</td>
</tr>
</tbody>
</table>

* SLN indicates speckled lentiginous nevi; CMN, congenital melanocytic nevus; NA, not applicable; and C/W, consistent with.
† At time of initial evaluation.
‡ Included portion of lesion that was macular “background” at age 6 months.

©2001 American Medical Association. All rights reserved.
Size, cm† | Type of Speckled Nevus | Histologic Features |
<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>6.5 x 3.0</td>
<td>Evolved from SLN to CMN with speckles</td>
<td>At age 6 months: compound nevus with some congenital features</td>
</tr>
<tr>
<td>3.0 x 4.0</td>
<td>Adjacent hybrid</td>
<td>At age 9 years: compound CMN‡ (Figure 2)</td>
</tr>
<tr>
<td>7.0 x 8.0</td>
<td>Adjacent hybrid</td>
<td>Compound CMN</td>
</tr>
<tr>
<td>5.9 x 5.0</td>
<td>Admixed hybrid</td>
<td>CMN: flat speckles (some junctional nests); raised portion (prominent dermal nests)</td>
</tr>
<tr>
<td>20.0 x 17.0</td>
<td>Admixed hybrid</td>
<td>NA</td>
</tr>
<tr>
<td>22.0 x 19.0</td>
<td>Admixed hybrid</td>
<td>NA</td>
</tr>
<tr>
<td>10.0 x 11.0</td>
<td>Admixed hybrid</td>
<td>NA</td>
</tr>
</tbody>
</table>

Entire diaper distribution Evolved from admixed hybrid to CMN with speckles CMN: focal atypia in a nodule

8.5 x 6.5 Evolved from admixed hybrid to CMN with speckles Compound CMN

20.0 x 10.0 Evolved from admixed hybrid to CMN with speckles Compound CMN; neurtization, sclerosis, and focal atypia of the dermal component in some portions; surrounded by a background area of melanocytic hyperplasia

REFERENCES


©2001 American Medical Association. All rights reserved.

Downloaded From:  on 10/16/2018