Segmental Speckled Lentiginous Nevus Exacerbated by Pregnancy in an Otherwise Healthy Female: A Case Report

Logan Kolb, DO,* Jessica Schweitzer, DO,* Mary Veremis-Ley, DO***

*Traditional Rotating Intern, Larkin Community Hospital, South Miami, FL
**Dermatology Resident, PGY-2, St. Joseph Mercy Hospital, Ann Arbor, MI
***Faculty, Department of Dermatology, St. Joseph Mercy Hospital, Ann Arbor, MI

Disclosures: None
Correspondence: Logan Kolb, DO; oganjkolb@gmail.com

Abstract

Speckled lentiginous nevi (SLN), or nevus spili, are seen in 0.2% to 2.3% of the population, presenting as tan patches with overlying hyperpigmented macules or papules in a speckled arrangement. Segmental SLN represent a small subset of SLN, with the segmental type comprised of larger, unilateral lesions that may rarely give rise to melanomas and have been reported to proliferate in response to ultraviolet light exposure. We present the case of a 40-year-old woman who presented with dark brown macules and papules scattered in a sharp, unilateral distribution on her back, chest, and abdomen. The lesions had been present for 10 years but had enlarged and darkened during her previous pregnancies. A clinical diagnosis of segmental speckled lentiginous nevus was made, and the patient was given appropriate instructions for sun protection and proper follow-up. This is the second reported case of segmental SLN with morphologic changes during pregnancy, suggesting an underlying hormonal component in its pathogenesis.

Introduction

Speckled lentiginous nevus (SLN), also referred to as nevus spilus, is a relatively common skin lesion, occurring in 2.3% of adults.[1] SLN lesions present as numerous, hyperpigmented macules and papules over tan-brown patches.[2] SLN has recently been categorized into macular and papular subtypes, known as nevus spilus maculosus and nevus spilus papulosus, respectively.[3] Segmental SLN are large lesions that present in a unilateral or zosteriform distribution. Such cases typically present at birth or in infancy and may worsen secondary to ultraviolet (UV) light exposure.[4] These have been reported to give rise to single or multiple melanomas, although this is rare.[4,5] In this case report, we present a 40-year-old female with adult-onset segmental SLN that worsened during pregnancy, review the disease's pathogenesis and management, and provide diagnostic pearls.

Case Report

A 40-year-old, otherwise healthy woman presented with a 10-year history of dark brown macules and papules on the right side of her torso. The lesions were asymptomatic but were noted to become darker and larger during past pregnancies. No previous workup or treatments had been attempted. A full review of systems, including visual changes, hearing loss, neurologic abnormalities, and musculoskeletal changes, was negative.

Physical examination revealed a female with Fitzpatrick type IV skin with a unilateral distribution of brown macules and papules in a speckled arrangement with background hyperpigmentation over the right side of the back, chest, and abdomen (**Figures 1, 2**). No epidermal nevi or port wine stains were noted. Due to the asymptomatic nature of the lesions and their clinical presentation, a clinical diagnosis of segmental speckled lentiginous nevus was made. The patient was educated on the importance of sun protection and close monitoring of her skin lesions with annual skin examinations.

Discussion

Despite continuing debate over the nosology of speckled lentiginous nevi (SLN) among melanocytic lesions, the etiology is likely multifactorial, with genetic and environmental factors playing a role.[6] Approximately 80% are present at birth or shortly after,[7] leading some authors to categorize SLN as a congenital melanocytic nevus, while others believe it to be an acquired process because the characteristic speckled hyperpigmentation may take years to develop.[6] Segmental or "zosteriform" presentations of SLN are also believed to be caused by somatic mosaicism, which consists of genetically distinct populations of cells that result from post-zygotic somatic mutations. Recently, activating mutations of HRAS were identified in smaller SLN lesions, whereas an activating missense NRAS mutation was identified in a case of congenital segmental SLN.[8,9]

Environmental factors also influence established SLN lesions. Ultraviolet (UV) light exposure has been reported to darken the lentiginous background hyperpigmentation and cause proliferation of the speckled macules and papules.[4] Additionally, in one case series, melanomas arising within segmental SLN occurred only in sun-exposed areas, which may be explained by epigenetic modification of tumor suppressor genes in lesional skin.[4] Our patient reported darkening and enlargement of her SLN during pregnancy, which to our knowledge has been reported in only one other case.[10] Of note, the other case of SLN worsened by pregnancy had a congenital onset, whereas our patient reported adult-onset of her SLN. There are similarities between the SLN in our patient and general characteristics of melasma, a hyperpigmentation disorder that commonly affects women's faces secondary to hormonal changes, including those associated with pregnancy or oral-contraceptive use. Melasma occurs more frequently in darker pigmented individuals (Fitzpatrick III-V), may be triggered by sun exposure and pregnancy – suggesting hormones may have played a role in the hyperpigmentation component our patient's SLN -- and has a genetic component, with 40% of affected patients reporting relatives also affected by the disease.[11]

A meticulous physical examination can give the clinician several important clues for the diagnosis of SLN and associated disorders. The underlying patch, a key finding in SLN, is absent in clinically similar disorders like agminated nevus and partial unilateral lentiginosis (also known as segmental lentiginosis).[5] Macular versus papular

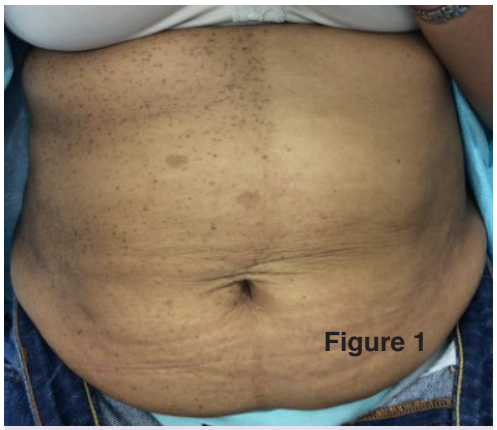

Figure 1. Dark brown macules and papules in a speckled, unilateral distribution overlying background hyperpigmentation with two associated café-au-late macules.

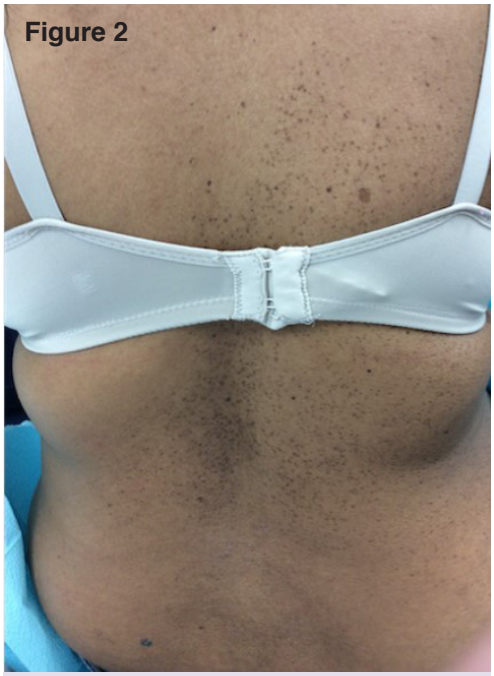

Figure 2. Dark brown macules and papules in a speckled distribution overlying background hyperpigmentation on the right side of the back.

presentations of SLN are also important clinical distinctions. Macular SLN are more likely to have an evenly spaced distribution, and they have a higher likelihood of malignant changes, whereas nevus spilus papulosis more often presents in an uneven, "star map" distribution and may be associated with phakomatosis pigmentokeratotica.[3] SLN have also been associated with phakomatosis pigmentovascularis (PPV) types III and IV[12] and speckled lentiginous nevus syndrome, which is characterized by SLN along with ipsilateral muscle weakness, hyperhidrosis, or sensory changes.[13] Type III PPV may be associated with multiple granular cell tumors.[14] Therefore, a thorough skin examination for port wine stains or nevus flammeus, an additional finding of PPV, should be performed. Other differential diagnoses to be considered for SLN-like lesions include café au lait macules, congenital nevi, lentigo simplex, and melanoma.

The diagnosis of SLN is often clinical, and management is similar to that of other large melanocytic nevi. On biopsy, the macules and papules are consistent with small junctional and compound nevi, respectively, with histology of background hyperpigmentation representing lentigo simplex.[2] The darkening of SLN lesions in response to UV exposure may be explained by these lentiginous features. Management of segmental SLN patients is similar to that of patients with large or numerous nevi. Patients should be followed with annual skin examinations and be educated on the importance of sun protection to prevent malignant changes within the lesion. Further treatment of lesions is often for cosmetic purposes and has been performed using Q-switched (QS) Nd:YAG, QS ruby, and QS alexandrite lasers.[15]

Conclusion

SLN typically present as smaller patches, though they may also have larger, segmental or zosteriform distributions. Such segmental cases are believed to be the result of somatic mosaicism affecting RAS proteins in the skin. SLN may worsen secondary to UV light exposure and have been reported to give rise to melanoma, which is more common in macular subtypes. We presented a case of segmental SLN in an otherwise-healthy, 40-year-old woman whose disease worsened during pregnancy; to our knowledge, this is only the second reported case in the literature of segmental SLN worsened by pregnancy.

References

1. Kopf AW, Levine LJ, Rigel DS, Friedman RJ, Levenstein M. Prevalence of congenital-nevus-like nevi, nevi spili, and café au lait spots. Arch Dermatol 1985;121:766-9.

2. Stewart DM, Altman J, Mehregan AH. Speckled lentiginous nevus. Arch Dermatol. 1978;114(6):895-6.

3. Vidaurri-de la Cruz H, Happle R. Two distinct types of speckled lentiginous nevi characterized by macular versus papular speckles. Dermatology 2006;212(1):53-8.

4. Ly L, Christie M, Swain S, Winship I, Kelly JW. Melanoma (s) arising in large segmental speckled lentiginous nevi: A case series. J Am Acad Dermatol. 2011;64(6):1190-3.

5. Torchia D. Segmentally grouped melanocytic nevi and melanoma risk. J Am Acad Dermatol 2012;66(2):324-5.

6. Schaffer J V, Orlow SJ, Lazova R, Bolognia JL. Speckled lentiginous nevus: within the spectrum of congenital melanocytic nevi. Arch Dermatol. 2001;137(2):172-8.

7. Lazova R. Speckled Lentiginous Nevus. Medscape [Internet]. Updated 19 Oct 2015 [cited 2 Mar 2016]. Available from: http://emedicine.medscape.com/article/1061990-overview.

8. Sarin KY, McNiff JM, Kwok S, Kim J, Khavari PA. Activating HRAS mutation in nevus spilus. J Invest Dermatol. 2014;134(6):1766-8.

9. Kinsler VA, Krengel S, Riviere JB, Waelchli R, Chapusot C, Al-Olabi L, Vabres P. Next-generation sequencing of nevus spilus-type congenital melanocytic nevus: exquisite genotype-phenotype correlation in mosaic RASopathies. J Invest Dermatol. 2014;134(10):2658-60.

10. Aloi F, Tomasini C, Pippione M. Agminated Spitz nevi occurring within a congenital speckled lentiginous nevus. Am J Dermatopathol. 1995;17(6):594-8.

11. Handel AC, Miot LDB, Miot HA. Melasma: a clinical and epidemiological review. An Bras Dermatol. 2014;89(5):771-82.

12. Fernández-Guarino M, Boixeda P, de las Heras E, Aboin S, García-Millán C, Olasolo PJ. Phakomatosis pigmentovascularis: clinical findings in 15 patients and review of the literature. J Am Acad Dermatol. 2008;58(1):88-93.

13. Happle R. Speckled lentiginous nevus syndrome. Delineation of a new distinct neurocutaneous phenotype. Eur J Dermatol. 2002;12:133–5.

14. Guiglia MC, Prendiville JS. Multiple granular cell tumors associated with giant speckled lentiginous nevus and nevus flammeus in a child. J Am Acad Dermatol. 1991;24(2):359-63.

15. Polder KD, Landau JM, Vergilis-Kalner IJ, Goldberg LH, Friedman PM, Bruce S. Laser eradication of pigmented lesions: a review. Dermatol Surg. 2011;37(5):572-95.