

CASE REPORT

A rare case report of a plaque type- blue nevus within a giant nevus spilus

Manish Rijhwani,¹ MD, Tanya Gupta,² Meet Patel,² Maneesh K Vijay,³ MD, Manisha Nijhawan,⁴ MD, Sankalp Awaṣṡhi,⁵ MD, Priyanka Sharma,² Pearl Dogra,²

¹Associate Professor, ²Resident, ⁴Professor & Head, ⁵Professor, Department of Dermatology

³Assistant Professor, Department of Onco-Pathology

ABSTRACT

Nevus spilus (NS), also known as speckled lentiginous nevus, is a variant of melanocytic nevus. It is characterized by a uniform light-brown patch within which there is speckling of darker macules or papules. Blue nevi are bluish-coloured lesions, formed by an intradermal proliferation of melanocytes. We describe a case of 29-year-old woman presenting with blue nevus in background of nevus spilus on the upper back and nape of neck.

INTRODUCTION

Nevus spilus, also known as speckled lentiginous nevus, is a condition characterized by hyperpigmented macules or papules scattered over a background of light tan brown pigmentation. The more pigmented smaller macules or papules are lentigines or melanocytic nevi, and the background diffuse pigmentation is a lentigo or a cafe au lait macule.^{1,2} Nevus spilus can be congenital, but is more commonly an acquired lesion. It has a benign behavior, though a few cases of melanoma arising on a nevus spilus have been reported.³ A cellular blue nevus (CBN) is defined as an aggregate of hamartomatous dermal pigmented melanocytes, typically occurring as a solitary lesion. It can be present at birth or acquired, and was originally described by the Swiss dermatologist Max Tièrehe in 1906.⁴ Blue nevi usually present as a single, acquired lesion, but very rarely can arise as multiple lesions grouped in a well-defined area, not usually larger than 10 cm

in diameter; they are then called agminated blue nevi.⁵ The combination of agminated blue nevi and nevus spilus is rare.⁶

CASE REPORT

A 29-year-old female, presented with complaint of an asymptomatic blackish patch on the nape of the neck and upper back since birth, which had been progressively increasing in size. There were no symptoms like itching, burning sensation or pain associated with the lesion. Examination revealed a single, well to ill-defined bluish black colored plaque of size 15 x 20 cm with prominent follicular openings overlying a well-defined, diffuse, hyperpigmented patch of size 40 x 35 cm present on upper back extending till the nape of neck (Fig. 1). Two biopsies were taken, one from the centre of bluish black plaque and second from the peripheral hyperpigmented patch. Histology of the first biopsy revealed a combined pattern with findings characteristic

Correspondence: Dr. Manish Rijhwani, Department of Dermatology



Fig. 1 Single, well to ill-defined bluish black colored plaque overlying a well-defined, diffuse, hyperpigmented patch on upper back extending till the nape of neck.

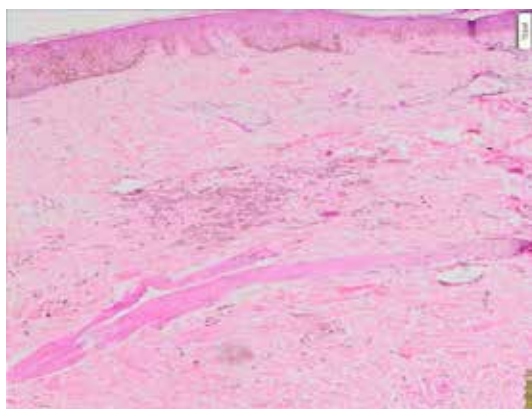


Fig. 2 Histology revealed dermal proliferation of spindle-shaped melanocytes associated with melanophages and lentiginous proliferation of melanocytes at the dermo epidermal junction and nests of melanocytes in the superficial dermis (H & E, original magnification X4).

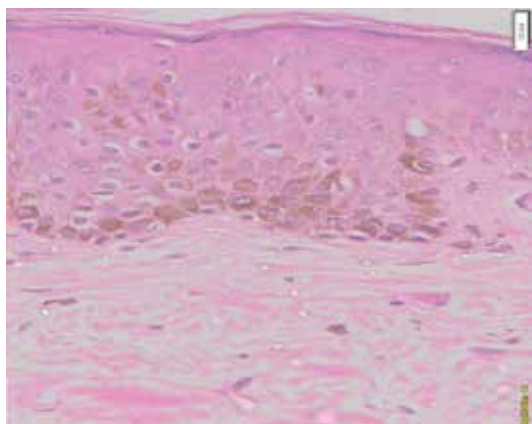


Fig. 3 Nevus spilus: lentiginous proliferation of melanocytes at the dermo epidermal junction and nests of melanocytes with no atypia in the superficial dermis. (H & E, original magnification X10).

of blue nevus (a dermal proliferation of spindle-shaped melanocytes associated with melanophages) and findings corresponding to nevus spilus (a lentiginous proliferation of melanocytes at the dermoepidermal junction and nests of melanocytes with no atypia in the superficial dermis) (Fig. 2). Together, these findings were consistent with a blue nevus within a nevus spilus. Histology of the second biopsy revealed a characteristic finding of nevus spilus (lentiginous proliferation of melanocytes at the dermo epidermal junction and nests of melanocytes with no atypia in the superficial dermis) (Fig. 3).

DISCUSSION

Cohen et al⁷ introduced the term nevus spilus for darkly spotted melanotic macules in 1970. In 1978, Stewart et al⁸ introduced the term speckled lentiginous nevus to describe darkly pigmented speckles on a tannish brown background. Since then, these two terms have been used interchangeably in the dermatology literature. Nevus spilus is a rare entity, with incidence estimated to be 0.2% to 2.3% among the general population.¹⁰ Cellular blue nevus (CBN) was first described by Tietze and Jaddosohn in 1906.⁴ This type of nevus is typically solitary and follows a relatively benign course. Phenotypes of blue nevus have been described, such as common, hypochromic, combined, 'polychromatic' (deep-penetrating), agminated 'plaque-type' and atypical blue nevi.¹¹ Blue naevi may appear in multiples, and if present in a well-circumscribed area of ≤ 100 mm, are considered an agminated blue nevus (ABN). The term 'agminate' is derived from the Latin word 'agminis' which means an army or a troop. ABN was first reported by Upshaw et al.⁴ Nevus spilus is thought to represent a localized

field defect and has been likened to “a garden of melanocytes” in which any type of nevus can develop, simultaneously or sequentially. Nevus spilus can be associated with blue nevus, with common melanocytic nevus, or with spitz nevus. The association of nevus spilus and agminated blue nevi is a rare combination, with few cases reported in the literature.^{6,9}

Kawamura presented a case and defined 3 histologic variants of this entity: type I, a combination of blue nevus and cellular nevus; type II, a combination of blue nevus with nevus spilus; and type III, a combination of blue nevus and fibromatous or myomatous nevoid formation. The cause of association between nevus spilus and blue nevus is unknown and may be attributed to a possible genetic susceptibility. The most common reported explanation is that of either an accidental overlap of two lesions, or that the combination of blue nevus is a clinical variant of nevus spilus.¹² The risk of malignant change is low in nevus spilus, although cases of melanoma arising on this pigmented lesion have been reported in the literature.¹³⁻¹⁵

CONCLUSION

Dermatologists need to be vigilant, as there have been reports of melanoma arising from within pre-existing nevus spilus, and therefore histological examination is important. Since blue nevus within a nevus spilus is a rarely reported entity, its prognosis is uncertain, making patient counselling challenging.

REFERENCES

1. Cruz HV, Happle R. Two distinct types of speckled lentiginous nevi characterized by macular versus papular speckles. *Dermatol* 2006; 212:53-58.
2. Grichnik JM, Rhodes AR, Sober AJ. Benign neoplasias and hyperplasias of melanocytes. In: Wolff K, Goldsmith LA, Katz SI, Gilchrist BA, Paller BS, Leffell DJ, eds *Fitzpatrick's Dermatology in General Medicine*, 7th ed. New York: McGraw-Hill, 2008:1099-1122.
3. Hoffman-Wellenhof R, Soyer HP, Smolle J, Kerl H. Spitz's nevus arising on a nevus Spilus. *Dermatol*. 1994; 189:265-68.
4. Upshaw BY, Ghormley RK, Montgomery H: Extensive blue nevus of Jadassohn-Tièche; report of a case. *Surg* 1947; 22:761-65.
5. Velez A, del-Río E, Martín-de-Hijas C, Furio V, Sánchez Yus E. Agminated blue nevi: Case report and review of the literature. *Dermatol*. 1993; 186:144-48.
6. Simonetti V, Grenzi L, Piana S, Albertini G, Longo C. Agminated blue nevus combined with nevus spilus: An uncommon association. *Int J Dermatol*. 2015;54:215.
7. Cohen HJ, Minkin W, Frank SB. Nevus spilus. *Arch Dermatol* 1970; 102:433-37.
8. Stewart DM, Altman J, Mehregan AH. Speckled lentiginous nevus. *Arch Dermatol* 1978; 114:895-96.
9. Kawamura T. Atypical blue nevus; report of a case. *Arch Dermatol*. 1950; 62:395-99.
10. Brito MHTS, Dionísio CSNM, Fernandes CMBM, Ferreira JCM, Rosa MJPMC, Garcia MMAPS. Synchronous melanomas arising within nevus spilus. *An Bras Dermatol*. 2017; 92(1):107-109.
11. Ferrara G, Soyer HP, Malvey J, Piccolo D, Puig S, Sopena J, Zalaudek I, Argenziano G: The many faces of blue nevus: a clinicopathologic study. *J Cutan Pathol* 2007; 34:543-51.
12. Marchesi L, Naldi L, Parma A et al. Agminate blue naevus combined with lentigo. A variant of speckled lentiginous nevus? *Am J Dermatopathol* 1993; 15:162-65.
13. Piana S, Gelli C, Grenzi L, Ricci C, Gardini S, Piana S. Multifocal melanoma arising on nevus spilus. *Int J Dermatol*. 2006; 45:1380-81.
14. Yoneyama K, Kamada N, Mizoquchi M, Utani A, Kobayashi T, Shinkai H. Malignant melanoma and acquired dermal melanocytosis on congenital nevus spilus. *J Dermatol*. 2005; 32: 454-58.
15. 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; 64:1190-93.