

# A giant congenital nevus spilus: A case report

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## ABSTRACT:

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Congenital nevus spilus is an uncommon condition and usually have small size.<sup>1</sup> It is found 0-0.2% of all newborns.<sup>2,8-12</sup> Nevus spilus is characterized by multiple pigmented macules or papules within a pigmented patch. Nevus spilus may occur anywhere on the body and have variable in size and configuration.<sup>3</sup>

We report a case of 5 year-old Thai girl with a large congenital nevus spilus extended from the left buttock to leg since birth, which is a rare condition and uncommon finding in congenital nevus spilus.

**Key word:** Nevus spilus, speckled lentiginous nevus, nevus on nevus, spotted nevus

## บทคัดย่อ :

ศิริณี วิภวกุล, จัตราพร แท่นชัยกุล, ปิ่นนรี ชัตติพัฒนาพงษ์ รายงานผู้ป่วยที่มาด้วยปานขนาดใหญ่ที่มีจุดเข้มเล็กๆ อยู่ภายใน (NEVUS SPILUS) ตั้งแต่เกิดที่บริเวณก้นจนถึงขาด้านซ้าย วารสารโรคผิวหนัง 2556; 29: 183-188.

สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

ปานสีไปรุส (nevus spilus) พบได้น้อยมากในทารกแรกเกิด พบได้ตั้งแต่ 0-0.2% และมักมีขนาดเล็ก มีลักษณะเป็น ปานสีน้ำตาลที่มีจุดเข้มเล็กๆ อยู่ภายใน พบที่ตำแหน่งใดก็ได้ มีขนาดและรูปร่างหลากหลาย รายงานฉบับนี้เป็นการนำเสนอ ผู้ป่วยเด็กหญิงอายุ 5 ปี มาด้วยปานขนาดใหญ่ที่มีจุดเข้มเล็กๆ อยู่ภายในตั้งแต่เกิด

**คำสำคัญ :** ปานแต่กำเนิด, ปานสีน้ำตาลที่มีจุดเข้มเล็กๆ อยู่ภายใน

## Introduction

Nevus spilus is a benign melanocytic condition which presents with numerous dark brown macules and papules within light tan patch. It is a common condition that is found in 2-3% of the population<sup>4</sup> but rare entity in newborn period.

In this report, we present a patient who presented with very large size nevus spilus since birth.

## Case report

A 5-year-old girl from Nongkhai was presented with a large light brown lesion on her left buttock and leg since birth. It gradually enlarged during childhood, and recently she noticed the development of multiple darkly pigmented papules within the large pigmented lesion. The patient was born full-term and healthy with neither illness nor other abnormalities. None of her family members was affected with a similar disorder.



**Figure 1** Multiple dark brown papules and nodules superimposed on a tan patch on left side extended from buttock to leg.

The skin examination revealed multiple, discrete, well-defined, smooth surface, dark brown papules and nodules superimposed on a tan patch size 40 x 10 cm on left side extended from buttock to leg (Figure1)

Histopathological examination from dark brown papules on left buttock, calf and hip showed compound melanocytic nevus with focal spitzoid feature (Figure2), intradermal and blue nevus (Figure3) and intradermal neves (Figure4), respectively. Furthermore, skin biopsy from tan patch on left calf showed basal hypermelanization (Figure5).

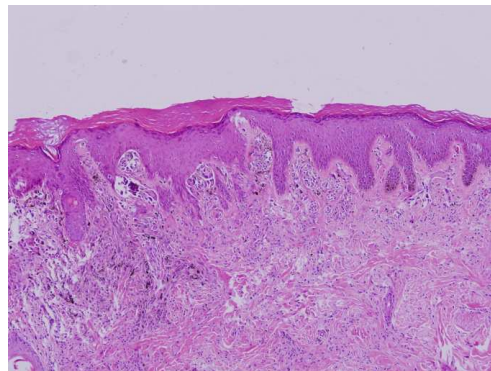


**Figure 2** Compound melanocytic nevus with focal spitzoid feature.

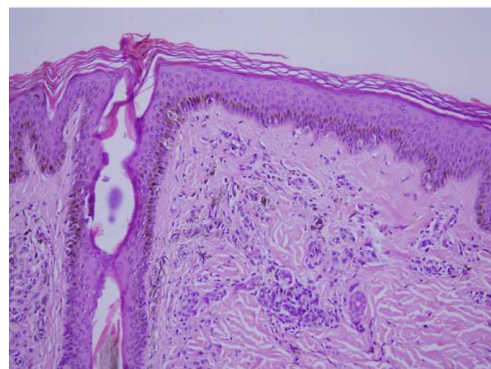


**Figure 3** Intradermal and blue nevus

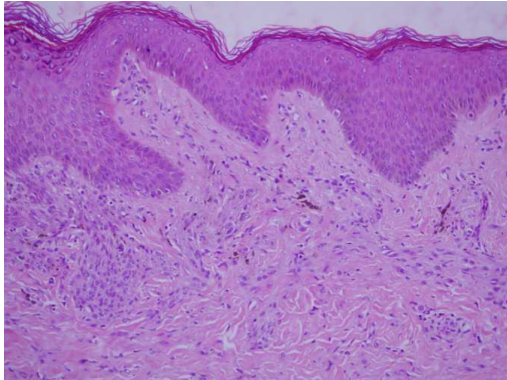


**Figure 4** Intradermal nevus

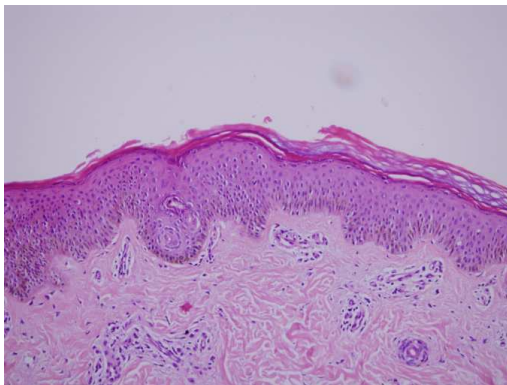


**Figure 5** Basal hypermelanization

Excisional biopsy of suspected lesion and regularly follow up for surveillance of new lesion that might be transformed to malignant melanoma is recommended.

## Discussion

Nevus spilus is also known as speckled lentiginous nevus, nevus on nevus, spotted nevus, spots on a spot, zosteriform lentiginous nevus and speckled nevus spilus.<sup>5-7</sup> The term “spilus” comes from “spilos”, which means “spot” in Greek.<sup>1,6</sup>

The prevalence of nevus spilus is age-dependent. Newborn infants have prevalence of nevus spilus varies from 0-0.2% in different studies.<sup>8-12</sup> The two large survey studies in 1058 and 4641 newborn infants did not detect nevus spilus lesion.<sup>9-10,12</sup> The prevalence is increasing to 1.2% to 2.1% in white school children and up to 2.3% in white adult, which means most lesions develop in the first year of life, and some lesions develop in childhood or adolescence.<sup>2,8,11</sup> There are no data in darkly pigmented persons. Males and females are equally affected.<sup>11</sup>

Nevus spilus is a benign condition. It presents as a solitary light brown macule or patch with no or few speckles. The speckles increase in number during childhood.<sup>1</sup> The background patch usually has diameter 2 to 10 cm. The speckles usually have diameter 1 to 3 mm, but may up to 9 mm.<sup>11</sup> It is classified in three types: small or medium size (<20cm), giant, and zosteriform.<sup>5-6,11</sup> It can occur anywhere, but commonly found on the trunk and lower limbs.<sup>6-7,11</sup>

The localized defect in neural crest melanoblasts in combination with genetic and environment factors is postulated as the pathogenesis of nevus spilus.<sup>2</sup> Nevus spilus may present as single clinical entity or in association with other congenital anomalies such as speckled lentiginous nevus syndrome (SLN), phacomatosis pigmentovascularis, phacomatosis pigmentokeratotic and FACES syndrome (unique facial feature, anorexia, cachexia and eye and skin abnormalities). SLN syndrome presents as nevus spilus

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of papular type combined with ipsilateral neurologic abnormalities such as dysesthesia, muscular weakness or hyperhydrosis.<sup>1,11</sup> Macular speckled lentiginous nevus is a hallmark of phacomatosis pigmentovascularis, as well as papular speckled lentiginous nevus which is found in phacomatosis pigmentokeratotic and SLN syndrome. Moreover, there is a report about the association between granular tumor and nevus flammeus with a giant nevus spilus.<sup>2</sup>

Nevus spilus carries a small risk of melanoma transformation.<sup>4-5</sup> The risk of developing melanoma in nevus spilus raises if it has been presented since birth, diameter more than 4 cm, and giant or zosteriform in morphology.<sup>11</sup> In a review of 25 cases of melanomas associated with nevus spilus which 60% of melanoma occurred on small or medium sized nevus spilus, 24% on zosteriform nevus spilus and 16% on giant nevus spilus.<sup>13</sup> Zosteriform and giant nevus spilus can be found in a very small number comparing to the total number of nevus spilus, these data indicate a high risk of melanoma development in these patients. Furthermore, the macular also has higher risk more than papular lesions.<sup>4</sup> The histological examination in majority of cases reveal superficial spreading melanoma.<sup>4</sup>

The histological findings of the background hyperpigmentation are represented in one of the other feature of a lentigo simplex, café-au-lait macule, melanocyte hyperplasia or nevus. The darker speckles are classically reported as junctional or compound

nevus, occasionally reported as intradermal, Spitz or blue nevus and rarely reported.<sup>4-6,8</sup>

Nevus spilus can rarely degenerate.<sup>12</sup> Until now, there is no standardized management for nevus spilus. Due to the small potential risk of developing melanoma, the patient with small or medium sized nevus spilus is advised to monitor and detect any changes by self-assessment. There is still controversy in prophylactic removal of the lesions which absence of any suspicious modification. Giant or zosteriform nevus spilus have higher risk developing melanoma and often difficult to accurately self examination, so the patients are indicated to closely follow up life long to exclude malignancy transformation.<sup>14</sup> The changes in size, color and elevation of nodules require biopsy.<sup>5</sup> Sequential digital dermoscope is an useful tool for early detection of melanoma. Efficacy of laser treatment is also controversy.

In conclusion, we report a case of giant congenital nevus spilus and treatment with biopsy few speckles for histopathology evaluation. Long term follow up is needed in this patient.

### References

1. Leung AK, Kao CP, Robson WL. A Giant Congenital Nevus Spilus in an 8-Year-Old Girl. *Adv Ther* 2006; 23: 701-4.
2. Kaur TD, Kanwar AJ. Giant nevus spilus and centrofacial lentiginosis. *Pediatr Dermatol* 2004; 21: 516-7.
3. Hanayama H, Terashi H, Hashikawa K, Tahara S. Congenital melanocytic nevi and nevus spilus have a tendency to follow the lines of Blaschko: an examination of 200 cases. *J Dermatol* 2007; 34: 159-63.
4. Prodinger C, Tatarski R, Laimer M, Ahlgrimm-Siess V. Large congenital nevus spilus-improved follow-up through the use of in vivo reflectance confocal microscopy. *Dermatol Pract Concept* 2013; 30: 55-8.

5. Hofmann-Wellenhof R, Soyer HP, Smolle J, Kerl H. Spitz's nevus arising on a nevus spilus. *Dermatology* 1994; 189: 265-8.
6. Fernandez-Flores A. Eponyms, morphology, and pathogenesis of some less mentioned types of melanocytic nevi. *Am J Dermatopathol* 2012; 34: 607-18.
7. Manganoni AM, Pavoni L, Farisoglio C, Sereni E, Calzavara-Pinton P. Report of 27 cases of naevus spilus in 2134 patients with melanoma: is naevus spilus a risk marker of cutaneous melanoma? *J Eur Acad Dermatol Venereol* 2012; 26: 129-30.
8. Rhodes AR. Nevus spilus: a potential precursor of cutaneous melanoma worthy of aggressive surgical excision? *Pediatr Dermatol* 1996; 13: 250-2.
9. Alper JC, Holmes LB. The incidence and significance of birthmarks in a cohort of 4,641 newborns. *Pediatr Dermatol* 1983; 1: 58-68.
10. Walton RG, Jacobs AH, Cox AJ. Pigmented lesions in newborn infants. *Br J Dermatol* 1976; 95: 389-96.
11. Vaidya DC, Schwartz RA, Janniger CK. Nevus spilus. *Cutis* 2007; 80: 465-8. Review.
12. Casanova D, Bardot J, Aubert JP, Andrac L, Magalon G. Management of Nevus Spilus. *Pediatr Dermatol*. 1996; 13: 233-8.
13. Corradin MT, Zattra E, Fiorentino R, Alaibac M, Belloni-Fortina A. Nevus spilus and melanoma: Case report and review of the literature. *J Cutan Med Surg* 2010;14:85-9
14. Cecchi R, Fancelli L, Troiano M. Melanoma arising in giant zosteriform nevus spilus. *Indian J Dermatol Venereol Leprol* 2012; 78: 643-5.