

Clinical, Dermoscopic and Histopathological Findings in Diagnosis of Nevus Spilus

Risa, A¹, Asri, E¹, Izrul I,² Tofrizal A³

¹ Dermato-Venereology Department of Andalas University/ Dr.M.Djamil Hospital, Padang, Indonesia

email: amillia.risa@yahoo.com

² Dermato-Venereology Department of Rasidin Hospital Padang Indonesia

³ Department of Anatomical Pathology Faculty of Medicine, Andalas University

Abstract

Introduction: Nevus spilus (NS) are seen in 0.2% to 2.3% of the population and have 0,13% to 0,2% risk for malignant transformation. Clinical, dermoscopic, and histopathological features were describe in this case report in order to be easily recognize NS. Although NS is a benign cutaneous anomaly it has potential malignant transformation and requires regular follow up. **Case Report:** A case of nevus spilus in 23 years-old female was reported. There were multiple asymptomatic brownish pigmented spots over brownish patch on left cheek which gradually increased in number and size since 1 year ago. Dermatologic state: brown macules and dark brown papules in a speckled, overlying background café au lait macule. Dermoscopy revealed reticular pattern in background light brown and dark reticuloglobular pattern in dark speckled. Histopathology showed elongation of rete ridges with grouping of melanocyte cells at the tip, and proliferation of nevus cells. **Conclusion:** Patient was planned to treat with Nd-Yag laser.

Keywords: dermoskopi, nevus on nevus, speckled lentiginous nevus, spotty nevus

Abstrak

Pendahuluan: Nevus spilus (NS) ditemukan sekitar di 0,2% hingga 2,3% dan memiliki resiko menjadi keganasan sekitar 0,13% to 0,2%. Gambaran klinis, dermoskopis, histopatologis dijelaskan dalam laporan kasus ini agar NS mudah dikenali. Walaupun NS adalah suatu lesi jinak namun memiliki postensi untuk menjadi keganasan sehingga dianjurkan untuk di follow up secara teratur. **Laporan Kasus:** Satu kasus nevus spilus telah dilaporkan pada pasien perempuan berusia 23 tahun. Terdapat beberapa bintik-bintik kecoklatan yang asimtomatik diatas bercak kecoklatan di pipi kiri yang secara bertahap meningkat dalam jumlah dan ukuran sejak 1 tahun lalu. Status dermatologis: makula kecoklatan dan papul kecoklatan diatas café au lait. Dermoscopy menunjukkan pola retikular dengan latar belakang coklat terang dan pola retikuloglobular gelap. Histopatologi menunjukkan perpanjangan rete ridges dengan pengelompokan sel melanosit di ujungnya, dan proliferasi sel nevus. **Kesimpulan:** Pasien direncanakan diterapi dengan laser Nd-Yag..

Kata kunci: dermoscopy, nevus on nevus, speckled lentiginous nevus, spotty nevus.

I. INTRODUCTION

Nevus spilus (NS), also known as speckled lentiginous nevus (SLN) occurs in approximately 1% to 2% of the population. They are present either at birth or in the first years of life and have therefore been regarded as a variant of congenital nevus. There does not appear to be gender or ethnic predilection. Darkly pigmented flat macules or papules are usually present within the nevus spilus on presentation. New pigmented elements may evolve within the lesion over time. The background pigmentation of a nevus spilus is circumscribed and similar in appearance to a café-au-lait macule in hue, with even light pigmentation. There are scattered superimposed more darkly pigmented macules or papules. The tan macular background pigmentation can range in size from less than 1 cm to larger than 10 cm in diameter. Lesions are most commonly found on the trunk and extremities, although any cutaneous site may be affected. Multiple lesions may be present and have a segmental distribution.

Dermoscopy reveals dark speckled foci with a reticuloglobular pattern in a background light brown and reticular pattern. Mixed patterns may occur that include combinations of homogeneous, reticular, globular, granular, and spitzoid patterns.^{1,2}

Histologic sections reveal that the tan background corresponds to a diffuse area where there is slight epidermal hyperplasia with basilar hyperpigmentation, from no perceptible increase in melanocytes to a slight increase in singular melanocytes along the basal layer. The dark spots correspond to small lentiginous junctional and compound nevi featuring increased numbers of single melanocytes and small nests along the DEJ and small, round melanocytes in the superficial dermis, in cases of compound nevi. The diagnosis of nevus spilus is usually straightforward.^{1,2}

No standard guidelines exist for the management of patients with nevus spilus. Clinical appearance (typical or atypical), history of stability or instability of pigmented elements, congenital or noncongenital onset, perceived risk of developing melanoma, and cosmetic concerns are considerations when determining whether to excise or recommend periodic clinical evaluation for life.³

II. CASE REPORT

A 23 year-old female, was referred to Dermato-Venereology out-patient Department of Dr. M. Djamil Hospital, with chief complaint multiple brownish pigmented spots on left cheek that not felt itchy or pain that gradually increased in number and size since 1 year ago. Initially there was brownish patch over the left cheek since birth that not felt itchy or pain. About 12 years ago there were multiple and small, brownish dots appeared over the patch which gradually increased in size and number, and become darker since 2 year ago. Patients never treated these complaints before. There is no history of yellow brownish verrucous patch on scalp, head or neck or another part of the body, increased sweating of the left half of her body. There is no history of muscular weakness, numbness, pain or discomfort within the involved skin and multiple brown macules on the another part of the body. There is no history of easy bleeding on brownish dots.

Physical examination we found within normal limit. Dermatologic state we found brown macules and dark brown papules in a speckled, overlying background café au lait macule (light brown macule), Fitzpatrick skin type IV. Routine blood examination were within normal.

Working diagnosed in this patient were nevus spilus, differential diagnosis were agminated acquired melanocytic nevi. We did dermoscopy examination and biopsy to establish the diagnosis.



FIGURE 1. A. SHOW THE LOCATION OF THE LESION IN LEFT CHEEK, UNILATERAL DISTRIBUTION. B. SHOW BROWN MACULES AND DARK BROWN PAPULES IN A SPECKLED, OVERLYING BACKGROUND CAFE AU LAIT MACULE.

Dermoscopy examination result.

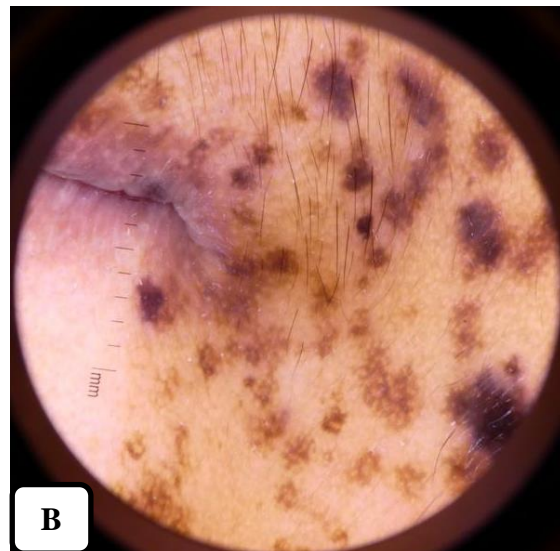
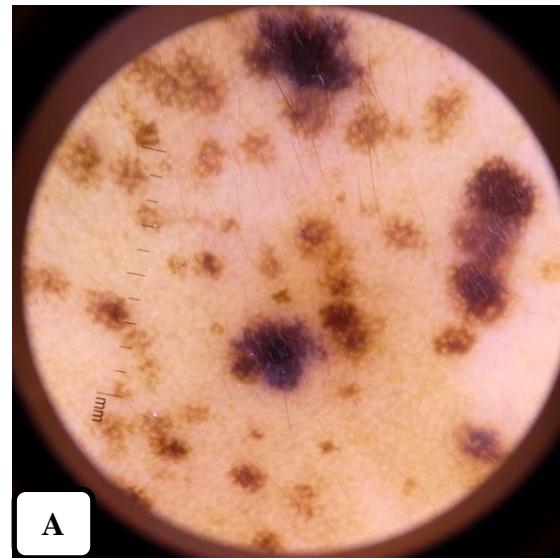


FIGURE 2. A&B DERMOSCOPY SHOWED DARKER BROWN AREA WITH A RETICULOGLOBULAR PATTERN IN DARK SPECKLED. THE BACKGROUND IS A LIGHT BROWN AND RETICULAR

Histopathological examination result were irregular acanthosis with elongation of rete ridges, a grouping of melanocyte cells at the tip of the rete ridges at several places, as well as increased pigmentation in the basal epidermis cells in epidermis. In Dermis, we can found proliferation of nevus, cells forming nests, especially in the upper epidermis and around the adnexa, with melanophag cells containing melanin pigments.

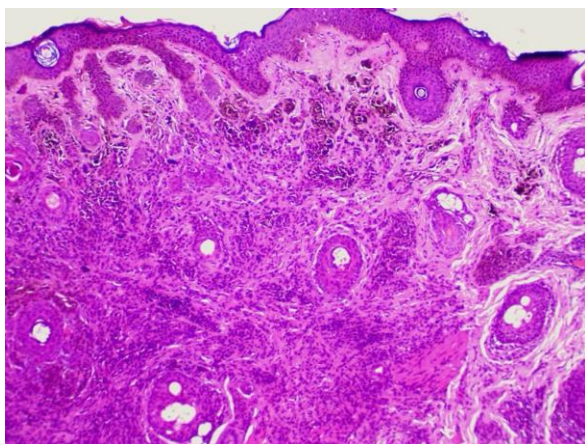


FIGURE 3. HISTOPATHOLOGICAL EXAMINATION SHOWED ELONGATION OF RETE RIDGES, PROLIFERATION OF NEVUS CELLS. (HE STAIN, MAGNIFICATION 10X)

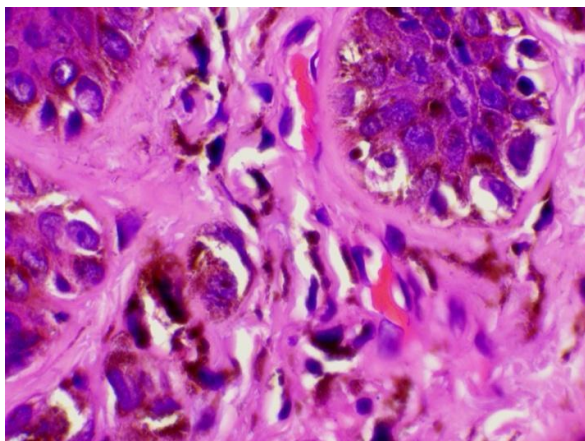


FIGURE 4. GROUPING OF MELANOCYTE CELLS AT THE TIP OF THE RETE RIDGES (HE STAIN, MAGNIFICATION 100X)

III. DISCUSSION

Nevus spilus also known as speckled lentiginous nevus, spots on a spot, and zosteriform lentiginous nevus. It is a rare dermatologic entity, occurring in less than 0.2% of newborn infants. The reported prevalence in the pediatric age group ranges from 1.3% to 2.1%, and that in the adult population is approximately 2.3%.^{4,5,6} The sex incidence is approximately equal. All ethnic and racial groups are at risk, but there is a slight predilection for white individuals. Presumably, nevus spilus results from a localized defect in neural crest melanoblasts under the influence of environmental and

genetic factors.³ In this case patient were 23 years old female.

Kamińska (Poland, 2003) reported 9 cases of nevus spilus, females were predominant, with an average age 37 years old.⁷ Ghosh et al (India, 2018) reported 7 cases of nevus spilus for 1 year in KPC medical hospital in Kolkata, females were predominant in this study. The most age of presentation was at birth and between 1-10 years old.⁸ Kamińska (Poland, 2003) reported 9 cases of nevus spilus, females were predominant, with an average age 37 years old.⁸ Handidwiono (Indonesia, 2015) reported 1 case of nevus spilus in a male patient, in a mental retardation elementary school/ among students with disabilities in Yogyakarta.⁹ Danarti et al (Indonesia, 2019) reported 2 cases of nevus spilus as a part of phacomatosis pigmentokeratotic.¹⁰ In an out-patient of dermatology venereology department in Dr. M. Djamil hospital, this is the first case that has been reported in the last ten years.

Nevus spilus can be congenital or acquired and usually appears during infancy but may also appear later in life, typically in the intensive growth stages.^{4,5} Aqil et al (Morocco, 2019) 70% of patients had these lesions from birth and 30% in adulthood.⁴ Kamińska (Poland 2003) reported, all patients present nevus at birth.⁷ In this case patient had lesion since birth.

Lesions are most commonly found on the trunk and extremities, although any cutaneous site may be affected. Multiple lesions may be present and have a segmental distribution.³ Kamińska (Poland 2003) reported lesions are commonly located on the extremities.⁷ Ghosh et al (India, 2018) reported majority of lesions were on the head and neck, and affected left side of the body, there are no bilateral lesions and associations with cutaneous or systemic findings.⁸ Aqil et al (Morocco, 2019) reported cases of nevus spilus with the most location were on the trunk.⁴ In this case patient had unilateral lesion on the face.

Typically, nevus spilus presents as an asymptomatic, tan to brown, sharply demarcated, well-circumscribed pigmentation that is studded with multiple, smaller, darker punctate macules or papules. The lesion often starts as a light-brown oval macule/patch with few or no speckles. Over time, the speckles slowly increase in size and number. These speckles can be macular or papular and represent lentigines or melanocytic nevi. The number of speckles usually ranges from 8 to 10, and the size of speckles ranges from 1 to 3 mm. The size of the background pigmented patch generally ranges from 2 to 10 cm. NS could develop more spot elements over the time. Nevus spilus, similar to melanocytic nevus, is classified as small (<1.5 cm), medium or intermediate (1.5– 19.9 cm), and large or giant (≥20 cm) according to their expected greatest diameter in adulthood. In most cases, the lesion is small or medium and solitary.^{6,11} In this case patient had the lesion since birth, then 12 years later ago there were multiple and small, brownish dots appeared over the patch which gradually increased in size and number, and become darker as the patients grew older. The classified of the lesion were medium.

Dermoscopy examination in this patient revealed darker brown area with a reticuloglobular pattern in dark speckled and light brown and reticular on the background. In line with Aqil et al (Morocco, 2019) also reported NS dermoscopy reveals darker brown areas with a reticuloglobular pattern. The background is usually light brown and reticular. In suspected cases of atypical NS, it sometimes reveals a hyperpigmented area with an irregular pattern.⁴ Kamińska (Poland, 2003) reported in all of the patients, the most typical pattern of dermoscopy observed was reticular, sometimes mixed with homogenous and globular compounds, located focally in the form of numerous patchy macules, varied in

color, ranging from light brownish to brown and blackish.⁷

Single lesion was excised for histological evaluation. The histopathological findings revealed irregular acanthosis with elongation of rete ridges. There is a grouping of melanocyte cells at the tip of the rete ridges at several places, as well as increased pigmentation in the basal epidermis cells in epidermis. In dermis revealed showed proliferation of nevus cells, cells forming nests, especially in the upper epidermis and around the adnexa, with melanophag cells containing melanin pigments. Vaidya (USA, 2007) reported, the light brown macule or patch usually shows mild melanocytic hyperplasia. Furthermore, it may show features of lentigo simplex, including small to moderate rete ridge elongation, increased melanocyte concentration in the basal layer, as well as increased melanin in both melanocytes and basal keratinocytes; and dermal melanophages. The speckles show junctional nests of nevus cells typically at the lowest points in the rete ridges, diffuse junctional activity, and dermal nevus cell aggregates.¹²

We make a differential diagnosed in this case is agminated acquired melanocytic nevi. Agminated acquired melanocytic nevi usually occur during puberty and lack background pigmentation. A skin biopsy revealed discrete nests of nevus cells at the dermoepidermal junction, mostly located on the accentuated tips of rete ridges. A markedly increased amount of melanin pigment was observed in the epidermis with melanophages in the superficial dermis. On dermoscopic examination, typical pseudo-network or globular pattern was seen. No background pigmentation within the agminated lesion.^{13,14} Therefore we consider that the present case is not compatible with agminated acquired melanocytic nevi.

No standard guidelines exist for the management of patients with nevus spilus.

Clinical appearance (typical or atypical), history of stability or instability of pigmented elements, congenital or noncongenital onset, perceived risk of developing melanoma, and cosmetic concerns are considerations when determining whether to excise or recommend periodic clinical evaluation for life. Documentation with high quality photographs can be used to aid follow up by parents and physicians.³ European Society of Laser in Dermatology recommended Q-switched laser for treated Nevus spilus.¹⁵ The Q-switched Nd:YAG laser emits a longer, near- infrared ray of 1,064 nm that is capable of penetrating into the deeper regions of the skin. Therefore, it is able to destroy deep-seated dermal melanocytes by selective photothermolysis.¹⁶ Patient were planned to treat with Q switched Nd-YAG laser.

Melanoma has been reported as a very rare complication in NS. The melanoma most often found on NS is superficial spreading melanoma, followed by nodular melanoma. Manganoni et al (Italy, 2011) evaluated 2134 patients with melanoma, 27 of them presented NS in a different body region. In the median 50 months of follow-up, patients were monitored with clinical and dermatoscopic observation. The risk of malignant melanoma in NS is higher in patients with the giant and/or zosteriform variants. Although in this case patient had a medium sized, we planing patient to follow up once per year, clinical and dermatoscopy examination is required to follow up the lesion. Patient also be instructed how to perform self examination to help detect early malignant changes. Dermatoscopy has been shown to improve the diagnostic accuracy for early melanoma detection. If there is any doubt, biopsy for histological evaluation should be considered. If a melanoma arises in an NS, its management and prognosis are similar to those for any other melanomas¹⁷⁻²⁰

IV. CONCLUSION

We reported a case of nevus spilus on facial, diagnosed was based on anamnesis, physical, dermoscopy and histopathological examination. Regular clinical-dermatoscopic observation is advised because of the risk of transformation into melanoma. Any changes lesion suspicious to malignant transformation should biopsied for histopathological evaluation.

V. REFERENCES

- [1] Corradin MT, Cacitti V, Giulioni E, Patriarca MM and Vettorello A. Nevus Spilus: A Review of the Literature. *SM Dermatolog J*. 2015; 1:1-7
- [2] Vaidya DC, Schwartz RA, Janniger CK. Nevus spilus. *Pediatric dermatology*. 2007;80:465-468
- [3] Cuda JD, Moore RF, Busam KJ. Melanocytic Nevi. In : Kang S, Amagai M, Bruckner AL, Enk AH, Margolis DJ, McMichael AJ, editors. *Fitzpatrick Dermatology*. Ed 9th. New York: McGraw Hill;2019:1944-1981
- [4] Aqil N, Elloudi S, Nassiri A, Gallouj S, Baybay H, Mernissi FZ. 2019. Dermoscopy of naevus spilus. *J Case Rept Img*. 2019. 1: 37-39
- [5] EL Jouari O, Senhaji G, El Mahi H, Gallouj S, Zahra MF. Dermoscopy of Nevus Spilus. *Madridge J Dermatol Res*. 2018; 3(2): 79-80
- [6] Leung AK, Barankin B. Circumscribed Pigmentation Stippled with Punctate Macules and Papules in a 9-year-old Boy. *Pediatrics in review*. 2018;39:e50-53
- [7] Kamińska W G. Dermoscopy of Nevus Spilus. *Dermatologic Surgery*. 2013; 39(10): 1550-1554.
- [8] Ghosh S, Ghosal L, Biswas SK, Bhunia D. A clinicoepidemiological study of different types of nevi in patients attending at a tertiary care hospital in Eastern India. *Journal of Pakistan Associations of dermatologists*. 2018; 28 (1); 51-58.
- [9] Handidwiono R, Budiyanto A, Radiono S, Rianawati T, Danarti R. Prevalensi kelainan kulit pada siswa sekolah dasar luar biasa (SDLB) pembina Yogyakarta. *MDVI*. 2015;42(3):119-127
- [10] Danarti R, Chusniyati N, Sulistiyowati Y. Phacomatosis pigmenkeratotica : two case series of a neurocutaneous rarity from Indonesia. *J Med Sci*. 2019;51(4): 309-316
- [11] Abrusci V, Benzecry V. Medium-sized nevus spilus of the neck treated with pulsed dye laser. *Dermatologic Therapy*. 2017;00:e12497.
- [12] Vaidya DC, Schwartz RA, Janniger CK. Nevus spilus. *Cutis*. 2007(80); 465-467
- [13] Shin J, Kim YC. Multiple agminated acquired melanocytic nevi. *Ann dermatol*. 2013;25(2); 251-252.
- [14] Shimasaki Y, Fukuta Y, Yoshida Y, Mori HH, Yamamoto O. Acquired agminated melanocytic

- naevi; report of two case and review of the literature. *Acta derm venereol.* 2012;92:585-663
- [15] Passeron T, Genedy R, Salah L, Fusade T, Kositratna G, Laubach J et al. Laser treatment of hyperpigmented lesions: position statement of the european society of laser in dermatology. *Journal of european academy of dermatology and venereology.* 2019;1-19
- [16] Kim YJ, Whang KU, Choi WB, Kim HJ, Hwang JY, Lee JH et al. Efficacy and Safety of 1,064 nm Q-switched Nd:YAG Laser Treatment for Removing Melanocytic Nevi. *Annals of Dermatology.*2012; 24(2), 162-167
- [17] 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? *Journal of the European Academy of Dermatology and Venereology.* 2011; 26(1): 129–130.
- [18] Corradin MT, Giulioni E, Fiorentino R, Santeufemia DA, Lo Re G, Vettorello A. In situ malignant melanoma on nevus spilus in an elderly patient. *Acta Dermatovenerologica Alpina, Pannonica, et Adriatica,* 2014, 23(1):17-19
- [19] Frings VG, Goebeler M, Kneitz H, *Dermpath & Clinic: In situ melanoma arising within a speckled lentiginous nevus.* *EJD.* 2018; 28.
- [20] Corradin MT, Zattra E, Fiorentino R, Alaibac M, Fortina AB. Nevus spilus and melanoma; case report and review the literature. *Journal of cutaneous medicine and surgery.* 2010 (14)2; 85-89