

NEVUS COMEDONICUS, A RARE CASE: DERMOSCOPIC AND HISTOPATHOLOGICAL FINDINGS

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ABSTRACT

Nevus comedonicus is an extremely rare adnexal hamartoma of pilosebaceous apparatus, with approximately 200 cases reported in the literature so far. It appears as a cluster of adjacent honeycomb-like dilated follicular openings with firm pigmented keratin plugs resembling comedones. The comedones are often arranged in a linear pattern parallel to Blaschko lines. We report a case of a 5-year-old boy with open brown to black comedones in a linear pattern localized on the back of the left thigh that appearing since birth. Pain, itch, and discharge did not obtain. However, there were some episodes of infection due to manual removal done by his mother, which left some hypertrophic scars. Dermoscopic examination revealed the distinctive pattern consisting of pigmented, sharply demarcated keratin plugs of 1-3 mm in diameter, some open pores, multiple structureless, various shades of brown homogenous circular areas surrounding the plugs. Histopathological examination showed an aggregation of dilated follicular infundibulum with laminated keratinous material plugging. This case report proves that dermoscopy examination, a simple, non-invasive diagnostic tool is very helpful in diagnosing nevus comedonicus. We recommend this tool to differentiate the diagnosis of other rare epidermal nevi, while histopathological examination should be performed only in uncertain cases.

Keywords: Dermoscopy; Epidermal Nevus; Nevus Comedonicus

INTRODUCTION

Nevus comedonicus is an extremely rare developmental abnormality of the pilosebaceous unit, which presenting as a constellation of dilated follicular orifices filled with dark keratinous plugs. It was first described as 'comedo nevus' by Kofmann in 1895.¹ Its prevalence has been estimated from 1 in 45,000 to 1 in 100,000, affecting any race and gender.² Retrospective study conducted in Mexico City from 1971 to 2001, among 417,511 pediatric patients, 443 displayed epidermal naevi, but only 5 of them diagnosed as nevus comedonicus.³ In Indonesia, there is no data on nevus comedonicus prevalence. The diagnosis of nevus comedonicus is relatively easy. It has a typical and different feature compared to other epidermal nevi. Dermoscopy, a safe, non-invasive easy-to-repeat diagnostic method mainly used in

Melanocytic lesion helps to rule out other differential diagnoses and establish the diagnosis of nevus comedonicus, making it beneficial, especially in pediatric cases.⁴ However, the use of this tool is rare, and only a few reports have been published regarding its use. A typical histopathological feature confirms the diagnosis in uncertain cases.

Here we present a rare case of nevus comedonicus with regard to its clinical, dermoscopic, histopathological features and the use of dermoscopy in this rare condition.

CASE REPORT

A 5-year-old Javanese boy presented to our clinic, Dermatovenereology Department dr. Moewardi General Hospital, Surakarta for evaluation of open brown to black, firm, and protrude comedones, localized to the back of his left thigh in a linear pattern along Blaschko lines (Figure 1) has occurred since birth. His mother reported that the lesions gradually

increased in number and size with time. The patient has ever been visited our department when he was one month old. At that time, he received various topical treatments, but there was no satisfactory improvement.

The parents retook him to our outpatient clinic after years due to their concern about this asymptomatic lesion which became increased in number and size. Again, there were no complaints of pain, itch, and discharge. His mother tried to remove the plugs herself, which left a big pore on the skin and caused some episodes of infections. Though it healed, it left some hypertrophic scars. Medical history was unremarkable, with no similar complaint in his family. The parents denied consanguinity. Pregnancy and labor were uneventful. The mother also denied any drug consumption during pregnancy.

Clinical examination on his back of the left thigh revealed multiple, comedo-like openings with brown to black, firm keratin plugs dispersed over a hypopigmented, linear spot of 20 x 4 cm, with some hypertrophic scars on it. His vital signs, body weight, and height were normal for his age. There were no other cutaneous findings on the other part of the body.



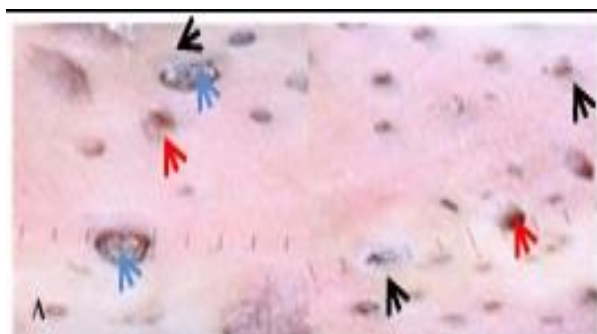
Figure 1. Nevus comedonicus on the back of the left thigh. A cluster of firm, pigmented, protuberant comedo-like papules in a linear pattern along Blaschko's line.

Ophthalmological and neurological examinations did not reveal any abnormality. Laboratory findings (complete blood count, liver functions, and kidney functions) were unremarkable. The dermoscopic revealed the distinctive pattern consisting of pigmented, sharply demarcated keratin plugs of 1-3 mm in

diameter, some open pores, multiple structureless, various shades of brown homogenous circular areas surrounding the plugs (Figure 2A). A 3 mm punch biopsy was obtained from the back of the left thigh. Histopathological examination showed an aggregation of dilated follicular infundibulum with laminated keratinous material plugging. The follicular walls were comprised of several layers of keratinocytes. The epidermis was within the normal limit (Figure 2B). This patient was diagnosed with nevus comedonicus based on the history taking, clinical examination which pictured the typical lesion of nevus comedonicus, dermoscopic and histopathological findings. He received topical therapy with emollient and 0.1% tretinoin gel. Marked improvement was seen a month after the treatment. The comedones had resolved, left some open pores and hypertrophic scars (Figure 3).

DISCUSSION

Nevus comedonicus is an extremely rare dermatological problem with an estimated 1 case in every 45,000–100,000 individuals.² A study by Inoue et al. reported that there were only 200 cases until the year of 2000.⁵ A retrospective study conducted in Mexico City from 1971 to 2001, among 417,511 pediatric patients, 443 displayed epidermal naevi. Still, only 5 of them diagnosed as nevus comedonicus.³ There has not been any data about the prevalence or even the case report of nevus comedonicus in Indonesia. Approximately 50% of nevus comedonicus cases appear at birth, with the other 50% developed symptoms during childhood, usually before the age of 10 years. There is no predilection for race or gender.⁶



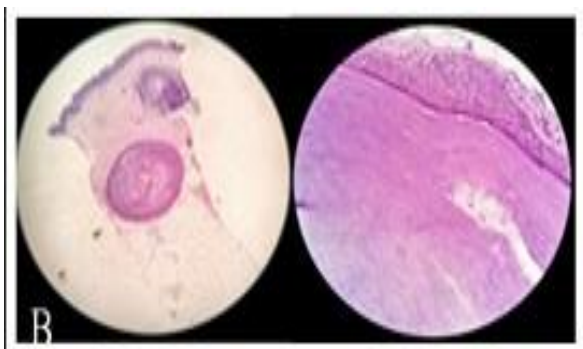


Figure 2A. Dermoscopy of nevus comedonicus. Multiple dilated follicular openings (red arrow) with pigmented keratin plugs 1-3 mm in diameter (blue arrow) and structureless, various shades of brown homogenous circular areas surrounding the plugs (black arrow) **B.** Histopathology of nevus comedonicus. Laminated keratinous material plugging on dermis and epidermis within normal limit. The follicular walls are comprised of several keratinocyte layers.



Figure 3A. Clinical appearance of the case prior to therapy. **B** Clinical appearance of the case a month after therapy with emollient and 0.1% tretinoin gel.

Clinically, nevus comedonicus present as a collection of discrete dilated follicular Ostia plugged with horny brown to black pigmented keratin. The lesions are most commonly found on the face, neck, upper arms, chest, and abdomen, usually arranged in groups, bands, or linear patterns along Blaschko lines.⁷ Normally, it is unilateral but can be bilateral in certain case.⁸ Nevus comedonicus is classified into two groups, reflecting the severity of the condition: the first group is characterized by the presence

of slightly pronounced skin lesions or comedo-like changes, which represent only a cosmetic defect, the second one presents with severe cutaneous symptoms including large cysts with scarring, often with a tendency to recurrence with the formation of fistulas and abscesses.⁹ Nevus comedonicus in unusual cases, may appear as an extensive inflammatory lesion involving large areas of the body, with inflammation and residual scarring.¹⁰

Several disorders have been known to be associated with nevus comedonicus. Cases showing any of these findings are included in nevus comedonicus syndrome, an entity considered within the larger group of epidermal nevus syndrome. Nevus comedonicus syndrome is characterized as a combination of nevus comedonicus with ocular defect (cataract, corneal erosion), skeletal defect (syndactyly, clinodactyly, preaxial polydactyly, absence of a ray of hand bones, scoliosis, vertebral defects), and neurologic defect (microcephaly, mental deficiency, dysgenesis of corpus callosum).¹¹ In our patient, the nevus comedonicus present alone without any other cutaneous or extracutaneous lesion, and also no abnormalities were found in the ophthalmological and neurological examination.

In our patient, the dermoscopic examination revealed the distinctive pattern consisting of dark, sharply demarcated keratin plugs of 1-3 mm in diameter, some open pores, numerous structureless, circular and barrel-shaped, homogenous areas with hyperkeratotic plugs of various shades of brown. These features were suggestive of nevus comedonicus. *Winciorek* and *Spiewak* defined dermoscopic features of nevus comedonicus as numerous circular and barrel-shaped homogenous areas in light and dark-brown shades with remarkable keratin plugs.¹² Dermoscopy as a diagnostic tool is a safe, non-invasive, and easy-to-repeat procedure mainly used in melanocytic lesions. It's also helpful in diagnosing nevus comedonicus.⁴ However, the use of this diagnostic tool has not been widely applied. To the author's best knowledge, supported by a thorough literature search (Pubmed), only four reports of the use of

dermoscopy in nevus comedonicus have been published.^{12,13,14,15} Dermoscopy is useful in differentiating nevus comedonicus from comedones of acne and other rare epidermal nevi, such as sebaceous nevus and hair follicle nevus. Comedones of acne vulgaris show numerous, homogenous areas, light, and dark-brown, sometimes black in color, depending on the type of acne, open or closed comedones, predominantly circular and situated superficially on dermoscopy. Sebaceous nevus shows bright, yellow spot which are not associated with hair follicles. Many follicular openings and interfollicular “pseudo-pigment network” on dermoscopy characterized hair follicle nevus.¹⁶

Histopathological examination of nevus comedonicus demonstrates a wide, deep invagination of the epidermis filled with keratin. These invaginations resemble dilated hair follicles; as evidence that they actually represent rudimentary hair follicles, occasionally found in the lower portion of an invagination one or even several hair shafts.¹⁷ Histopathological findings in our patient are in line with those previously reported. Histologically it is important to differentiate it with comedonal acne. In comedonal acne, the pilosebaceous units are complete, whereas those in nevus comedonicus are poorly formed. Furthermore, in nevus comedonicus, *hyperkeratosis* and *papillomatosis* are frequently seen in the interpapillary epidermis and absent in comedonal acne. The dilated pore of Winer can sometimes be confused with nevus comedonicus histologically. However, this condition is usually observed in the elderly and can be differentiated clinically.

Clinical findings themselves can be used to establish the diagnosis of nevus comedonicus as the diagnosis of nevus comedonicus is predominantly clinical. The differentiation of nevus comedonicus from other epidermal nevi is easy as the former shows the presence of “comedones”, which on extraction will leave a big pore on the skin surface. The finding of groups of lesions paralleled Blascko’s lines ruled out comedonal acne. In most cases, dermoscopy may prove helpful, while the biopsy is only indicated in uncertain cases.

CONCLUSION

Diagnosing nevus comedonicus is relatively easy. However, clinicians should be aware of the potential association of nevus comedonicus with other cutaneous lesions and extracutaneous features, such as ocular, skeletal and neurologic abnormalities as nevus comedonicus syndrome. Furthermore, our case report shows a typical feature and dermoscopic findings of nevus comedonicus which is rarely described in the literature. This case report also proves that dermoscopy examination, a simple non-invasive diagnostic tool is very helpful in diagnosing nevus comedonicus. We recommend this tool to differentiate the diagnosis of other rare epidermal nevi, such as sebaceous nevus and hair follicle nevus. At the same time, a histopathological examination should be performed only in uncertain cases.

REFERENCES

1. Kofmann S. A case of rare localization and spreading of comedones. *Arch Dermatol Syphilol.* 1895;32:177-8.
2. Kaliyadan F, Bhimji S. *Nevus Comedonicus.* Treasure Island: StatPearls Publishing; 2017.
3. Vidaurri-De La Cruz H, Tamayo-Sánchez L, Durán-McKinster C, Orozco-Covarrubias MDLL, Ruiz-Maldonado R. Epidermal nevus syndromes: Clinical findings in 35 patients. *Pediatr Dermatol.* 2004;21(4):432-9.
4. Kamińska-Winciorek G, Spiewak R. [Basic dermoscopy of melanocytic lesions for beginners]. *Postepy Hig Med Dosw (Online).* 2011;65:501-8.
5. Inoue Y, Miyamoto Y, Ono T. Two cases of nevus comedonicus: Successful treatment of keratin plugs with a pore strip. *J Am Acad Dermatol.* 2000;43(5 SPEC. SUPPL.):927-9.
6. Pierson T, Bandel C, Ehrig T. Benign epidermal tumors and proliferations. In: *Dermatology.* London: Mosby;2012:h.1795-1815.
7. Solomon LM, Esterly NB. Epidermal and other congenital organoid nevi. *Curr Probl Pediatr.* 1975;6(1): 3-56.
8. Mahran AM, Abdelsamea GM, Mekkawy

- MM. Bilateral Nevus Comedonicus Successfully Treated with Topical Adapalene: A Case Report. *Clin Dermatology Open Access J.*2017;2(6):000136.
9. Guldbakke KK, Khachemoune A, Deng A, Sina B. Naevus comedonicus: A spectrum of body involvement. *Clin Exp Dermatol.*2007;32(5):488-92.
 10. Kirtak N, Inaloz HS, Karakok M, Erguven HG, Ozgoztasi O. Extensive inflammatory nevus comedonicus involving half of the body. *Int J Dermatol.*2004;43(6):434-6.
 11. Happle R. The group of epidermal nevus syndromes. Part I. Well defined phenotypes. *J Am Acad Dermatol.*2010;63(1):1-22.
 12. Kamińska-Winciorek G, Śpiewak R. Dermoscopy on nevus comedonicus: A case report and review of the literature. *Postep Dermatologii i Alergol.* 2013;30(4):252-4.
 13. Vora R, Kota R, Sheth N. Dermoscopy of nevus comedonicus. *Indian Dermatol Online J.* 2017;8(5):388.
 14. Kayiran MA, Cebeci F, Zindanci I, Gurel MS, Akdeniz N. *North Clin Istanbul.* 2019;6(4):401-2.
 15. Chauhan P, Dilip M, Sharma S. Dermoscopy of nevus comedonicus: A case series. *IJPD.* 2020;21(3): 233.
 16. Okada J, Moroi Y, Tsujita J, et al. Hair follicle nevus - A dermoscopic approach. *Eur J Dermatology.* 2008;18(2):185-7.
 17. Elder D, Elenitsas R. *Lever's Histopathology of the Skin, 10th Edition.* Vol 36. Lippincott Williams & Wilkins; 2009.