



ORIGINAL ARTICLE

Dermoscopic Features of Small, Medium, and Large-Sized Congenital Melanocytic Nevi

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Background: Congenital melanocytic nevi (CMN) are present at birth. It is well known that the presence of large-sized congenital nevus in early life could predict a major risk of developing melanoma. **Objective:** To investigate the clinical and dermoscopic features of the CMN, to search for and highlight any differences between small-sized, medium-sized, large-sized CMN. **Methods:** A nonrandomized observational study was performed. A total of 108 melanocytic nevi were analysed by clinical and dermoscopic examination. **Results:** Of the subjects, 57.4% were aged less than 16 years, 42.6% were aged 16 and more. Of the nevi, 26 had reticular pattern (24.1%), 35 had globular pattern (32.4%), 13 had reticular-globular pattern (12.0%), 16 had homogeneous pattern (14.8%), 6 had reticular-homogeneous pattern (5.6%), 2 had globular-homogeneous pattern (1.9%), 7 had cobblestone pattern (6.5%), 3 had reticular patchy pattern (2.8%). Atypical dots and globules, focal hypopigmentation and perifollicular hypopigmentation are the most common dermoscopic features of CMN. The rarest dermoscopic feature is the blue-whitish veil. **Conclusion:** Most of the dermoscopic features related with dysplastic nevi up to the present, such as atypical dots and globules, focal hypopigmentation, perifollicular hypopigmentation were observed in CMN, in our study. Congenital nevus and dysplastic nevi may share the

same dermoscopic features, therefore it is important to know it is found at birth or not. (*Ann Dermatol* 29(1) 26 ~ 32, 2017)

-Keywords-

Dermoscopy, Nevus

INTRODUCTION

Congenital melanocytic nevi (CMN) are defined as nevi present at birth or appearing during subsequent months¹. CMN are present at birth on the skin of 1% to 6% of newborns and are usually classified according to their size as small (< 1.5 cm), medium (1.5 ~ 19.9 cm), and large/giant (> 20 cm)²⁻⁵. Most of the CMN are smaller than 1.5 cm³. CMN may not exist at birth, because of the embryonic absence of pigmentation and may appear clinically after the pigment developments months to years after birth^{2,6,7}. Furthermore, these lesions are described as 'congenital nevus-like nevi' or 'tardive congenital nevi' by some authors^{2,6}. The estimated prevalence of CMN ranges from 0.5% to 31.7%⁸. Small CMN have an estimated incidence of 1 in 100 live births, whereas large CMN is 1 in 20,000 live births⁹⁻¹¹. They occur in all races and ethnic groups, and males and females have an equal risk. Many of the nevi termed "dysplastic" or "atypical" are actually small CMN¹². CMN may develop due to a mutation that affects the morphogenesis of the embryonic neuroectoderm and migration of precursor cells to the skin.

Our aim was to investigate the dermoscopic patterns of CMN among study participants and to assess the dermoscopic patterns associated with the size of nevi as well.

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MATERIALS AND METHODS

A total of 108 patients attending the Department of Dermatology at Bezmialem Vakif University Hospital in Istanbul, a 2-year period were recruited for the study. Essential inclusion criteria were the presence of at least one nevus and the ability of the participants to state with certainty whether each nevus was present at birth or appeared in the first 2 years of life. Lesions which were located on mucosal, subungual, and acral sites were excluded from our study.

A total of 108 melanocytic nevi in 108 consecutive Caucasian patients were evaluated. After verbal consent was obtained from each patient, all study participants received clinical and dermoscopic examinations. Clinical data were obtained for each patient and included the following: sex, age, skin phototype, topography, diameter, color, dermoscopic pattern, symmetry, borders, atypical pigment network, hypertrichosis, perifollicular hypopigmentation, milia-like cysts, radial streaks, regression, atypical dots/globules, structureless areas, and blue-whitish veil.

Dermoscopic images of all the lesions were acquired at a 30× magnification and were stored in a digital imaging system (Fotofinder, Digital Dermoscopy; Foto Finder Systems GmbH, Bad Birnbach, Germany). All digital images were examined by the same dermatologist and were evaluated for global and local features. The size of the nevus was calculated by using specific software.

Dermoscopic patterns were classified as reticular, globular, cobblestone, reticuloglobular, homogeneous, peripheral reticular with a central homogeneous area, peripheral globular with a central homogeneous area, and reticular patchy.

CMN were divided into 3 groups, according to the diameter of the lesion: Small-sized congenital nevus is defined as having a diameter less than 1.5 cm; medium-sized congenital nevus is defined as having a diameter more than 1.5 cm but less than 20 cm; large congenital nevus is defined as having a diameter more than 20 cm.

Statistical analysis

Descriptive analysis of the sample was performed, including percentages for categorical variables, and mean and standard deviations for continuous variables. Comparisons between categorical variables were performed with chi-square tests and Fisher corrections when required. $p < 0.05$ was considered significant.

RESULTS

Clinical and dermoscopic examinations were performed on 108 participants.

Descriptive results

Sixty-two participants (57.4%) were aged less than 16 years, and 46 participants (42.6%) were aged 16 and more. Table 1 provides all patient demographic data.

There were 52 small-sized nevi (48.1%), 49 medium-sized nevi (45.4%), and 7 large-sized nevi (6.5%). Thirteen small-sized nevi (25.0%) were located on the extremities, 31 (59.6%) on the trunk, and 8 (15.4%) on the face and neck area. Twelve medium-sized nevi (24.5%) were located on the extremities, 29 (59.2%) on the trunk, and 8 (16.3%) on the face and neck area. Two large-sized nevi (28.6%) were located on the extremities, 4 (57.1%) on the trunk, and 1 (14.3%) on the face and neck area. No differences were observed between the location of nevi and size ($p=0.999$).

Twenty patients with Fitzpatrick skin type III (38.5%) and 32 patients (61.5%) with Fitzpatrick skin type IV had small-sized congenital nevi. One patient (2.0%) with Fitzpatrick skin type II, 13 (26.5%) with Fitzpatrick skin type III, and 35 (71.4%) with Fitzpatrick skin type IV had medium-sized congenital nevi. Two patients with Fitzpatrick skin type III (28.6%) and 5 (71.4%) with Fitzpatrick skin

Table 1. Demographic data of patients (n=108)

Characteristic	Value
Age (yr)	
Range	6~55
Median	24
Mean	26.34±6.71
Sex	
Male	51 (47.2)
Female	57 (52.8)
Sunburns	
Never	15 (13.9)
Mild~moderate	82 (75.9)
Severe	11 (10.2)
Fitzpatrick skin type	
II	1 (0.9)
III	35 (32.4)
IV	72 (66.7)
Location	
Face and neck	17 (15.7)
Trunk	64 (59.3)
Extremity	27 (25.0)

Values are presented as number only, mean±standard deviation, or number (%).

type IV had large-sized congenital nevi.

Dermoscopic patterns

1) General dermoscopic structures in congenital nevi

On dermoscopic examination, we found the presence of these patterns: reticular pattern, 24.1% (n=26); globular pattern, 32.4% (n=35); reticular-globular pattern, 12% (n=13); homogeneous pattern, 14.8% (n=16); reticular-homogeneous pattern, 5.6% (n=6); globular-homogeneous pattern, 1.9% (n=2); cobblestone pattern, 6.5% (n=7); reticular patchy pattern, 2.8% (n=3) (Fig. 1~4, Table 2). The globular pattern as the predominant dermoscopic pattern was more frequent in children younger than 16 years old (32.4%); the reticular pattern as the pre-

dominant pattern was more frequent in adults older than 16 years old (28.3%).

Perifollicular hypopigmentation was present in 32 congenital nevi (29.6%). An atypical pigment network was observed in 30 congenital nevi (27.8%). Milia-like cysts were present in 23 congenital nevi (21.3%). Hypertrichosis was observed in 17 congenital nevi (15.7%). Radial streaks were observed in 15 congenital nevi (13.9%). Focal hypopigmentation was present in 70 congenital nevi (64.8%). Atypical dots/globules were observed in 72 congenital nevi (66.7%). Regression was present in 6 congenital nevi (5.6%). Blue-whitish veil was present in 3 congenital nevi (2.8%). Vascular structures were always absent.



Fig. 1. Globular pattern with central dark globules.

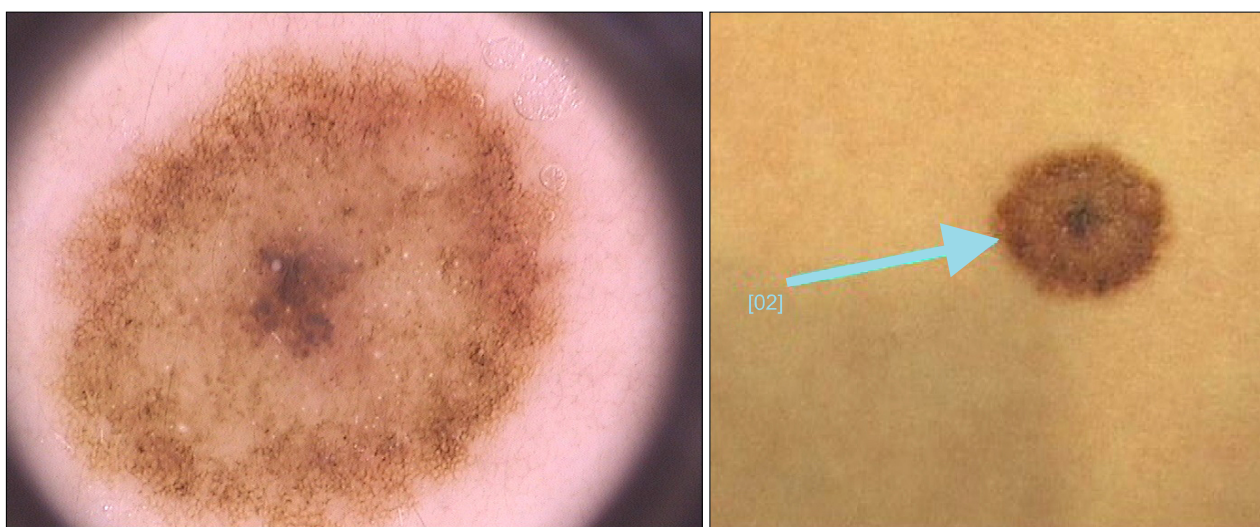


Fig. 2. Peripheral reticular pattern with central homogen area.



Fig. 3. Globular pattern with hypertrichosis.

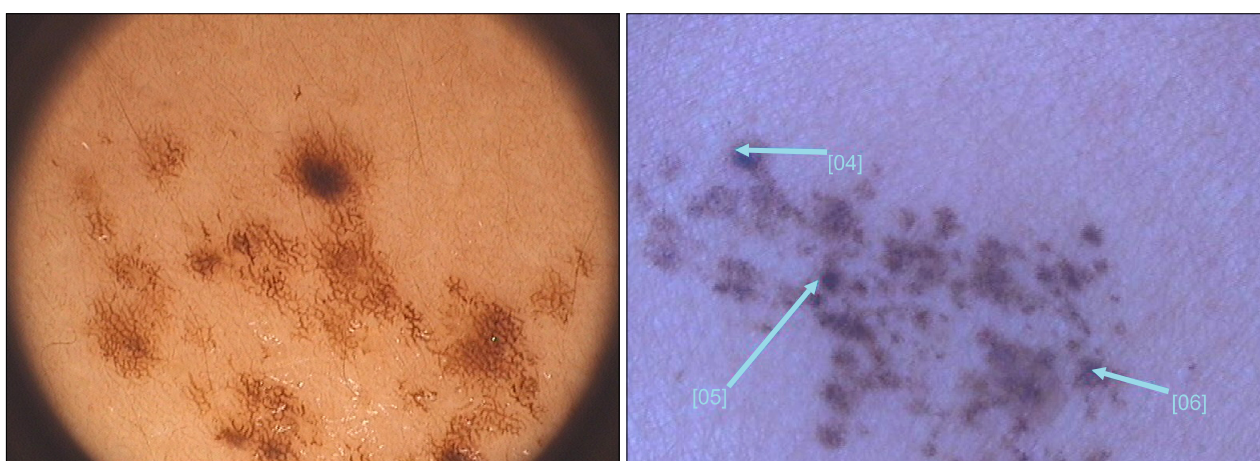


Fig. 4. Patchy reticular pattern.

Table 2. Dermoscopic features according to the size of the nevus

Dermoscopic features	Small-sized (n=52)	Medium-sized (n=49)	Large-sized (n=7)
Border regularity	19 (36.5)	17 (34.7)	2 (28.6)
Perifollicular hypopigmentation	15 (28.8)	14 (28.6)	3 (42.9)
Milia-like cysts	6 (11.5)	16 (32.7)	1 (14.3)
Hypertrichosis	2 (3.8)	11 (22.4)	4 (57.1)
Radial streaks	5 (9.6)	9 (18.4)	1 (14.3)
Focal hypopigmentation	29 (55.8)	38 (77.6)	3 (42.9)
Atypical dots and globules	26 (50.0)	39 (79.6)	7 (100)
Regression	3 (5.8)	1 (2.0)	2 (28.6)

Values are presented as number (%).

2) Relation between the size and dermoscopic structures in congenital nevi

On dermoscopic examination, we found the presence of these patterns: reticular pattern, 34.6% (n=18); globular pattern, 36.5% (n=19); homogeneous pattern, 19.2%

(n=10); reticular-homogeneous pattern, 5.8% (n=3); and cobblestone pattern, 3.8% (n=2) in small-sized congenital nevi; reticular pattern, 14.3% (n=7); globular pattern, 30.6% (n=15); reticular-globular pattern, 22.4% (n=11); homogeneous pattern, 12.2% (n=6); reticular-homogeneous pattern, 6.1% (n=3); globular-homogeneous pat-

tern, 4.1% (n=2); cobblestone pattern, 6.1% (n=3); reticular patchy pattern, 4.1% (n=2) in medium-sized congenital nevi; reticular pattern, 14.3% (n=1); globular pattern, 14.3% (n=1); reticular-globular pattern, 28.6% (n=2); cobblestone pattern, 28.6% (n=2); reticular patchy pattern, 14.3% (n=1) in large congenital nevi.

No association was found between the border regularity ($p=0.911$), perifollicular hypopigmentation ($p=0.744$), radial streaks ($p=0.442$), regression ($p=0.072$), blue-whitish veil ($p=0.543$) and size. Medium-sized congenital nevi had a statistically higher ratio of milia-like cysts ($p=0.03$) and focal hypopigmentation ($p=0.031$) than the others.

An atypical pigment network was observed in all of the large-sized congenital nevi (100%), 18 (36.7%) medium-sized congenital nevi, and 5 (9.6%) small-sized congenital nevi. There was a statistically significant difference related to the atypical pigment network between groups ($p=0.025$). Atypical pigmentation is a common dermoscopic feature in large-sized and medium-sized congenital nevi.

Hypertrichosis was observed in 2 (3.8%) small-sized nevi, 11 (22.4%) medium-sized congenital nevi, and 4 (57.1%) large-sized congenital nevi. There was an association and a highly positive correlation between hypertrichosis ($r=0.485$, $p=0.000$), the presence of atypical dots/globules ($r=0.783$, $p=0.001$), asymmetry ($r=0.552$, $p=0.006$) and the size of the lesion.

No association was found between asymmetry ($p=0.975$) border regularity ($p=0.353$), perifollicular hypopigmentation ($p=0.084$), radial streaks ($p=0.579$), regression ($p=0.407$), focal hypopigmentation ($p=0.243$) and the location of nevi.

DISCUSSION

Congenital melanocytic nevi (CMN) are described as neural crest-derived hamartomas, which appear at, or shortly after birth as pigmented tumors, by some authors⁸. The incidence of CMN in neonates ranges from 0.2% to 2.1% regardless of nevus size¹³. It is known that the large-sized CMN have a higher risk of malignant melanoma (MM) development than small- and medium-sized nevi. MM arising in CMN usually develops at younger ages and is located deeper within large CMN and superficially within small- or medium-sized CMN. Two important meta-analyses have been conducted to determine the significance of MM in CMN^{14,15}. In one of these reviews, 10 of the 432 patients developed melanoma within their giant congenital nevi. Krengel et al.¹⁵ analyzed 14 studies with a total of 6,571 patients CMN with who were followed for a mean of 3.4~23.7 years. Forty-six (0.7%) developed a to-

tal of 49 melanomas (mean age at diagnosis: 15.5 years, median age: 7 years). The authors found a markedly increased risk of developing melanoma during childhood and adolescence¹⁵. Large CMN are sometimes associated with neurocutaneous melanocytosis (NCM). NCM is a melanocytic proliferation including the cranial nervous system; this can increase intracranial pressure. In our study, none of the patients with large CMN had the symptoms of intracranial pressure. Brain MRI examinations were normal.

It has been estimated that the lifetime risk of developing melanoma is 1 in 100 for patients with small and medium CMN^{16,17}.

Congenital melanocytic nevi should be managed conservatively if there are no abnormalities at clinical and dermoscopic examination and the patient has no cosmetic disturbance¹⁸. Annual clinical and digital dermoscopic examination is indicated in these patients. Therefore, the physician should know the dermoscopic features of CMN to avoid unnecessary excisions, and to recognize melanoma when it begins to develop. In a nationwide study, melanoma developed in 3 of 131 (2.3%) patients with giant CMN¹⁸. Diagnosis of large CMNs is simple, regarding their size and appearance since birth¹⁹. Therefore, we did not include so many large CMNs in our study. We aimed to identify dermoscopic features of small- and medium-sized CMN, especially.

In most cases, dermoscopic examination reveals a globular or cobblestone pattern in large CMN. In our study, reticular-globular and cobblestone patterns were the most common patterns of large CMN in line with findings in the literature. The predominant dermoscopic patterns in small-sized and medium-sized nevi were globular, present in 36% of the small-sized group, and in 30% of the medium-sized group. Our results are consistent with those of previous studies^{20,21}. Additionally, the predominant dermoscopic pattern was globular pattern in patients younger than 16 years old, whereas it was reticular pattern in patients older than 16 years. Some authors suggest that a predominantly reticular pattern is present particularly in individuals 12 years or older^{22,23}. Our results support this finding.

In general, atypical dots/globules, focal hypopigmentation, and perifollicular hypopigmentation are the most common dermoscopic features of CMN found in our study. Our results are consistent with the results of Stinco et al.²⁴. Atypical dots/globules, focal hypopigmentation, asymmetry, and border regularity were common in both of the children and adults.

Milia-like cysts, hypertrichosis, radial streaks were uncommon than atypical dots and globules, focal hypopig-

mentation and perifollicular hypopigmentation, in general. The rarest dermoscopic feature was regression, according to our study.

The presence of atypical dots/globules, asymmetry, and hypertrichosis correlate with the size of CMN. It is known that these features are typical for giant and large CMN, and our results are consistent with findings in the literature²⁵. Focal hypopigmentation and milia-like cysts are statistically more common in medium-sized nevi than small-sized and large-sized nevi. There is no data about the rates of focal hypopigmentation and milia-like cysts, according to the size of nevi reported previously in the literature. Regression and radial streaks are the rarest dermoscopic features in all sizes of nevi. Perifollicular hypopigmentation and border regularity are other common features in all sizes of nevi without a statistical difference.

Small-sized nevi are primarily located on the trunk (59.6%), and the most frequent dermoscopic features in small-sized nevi are focal hypopigmentation (55.8%) and atypical dots/globules (50.0%). Medium-sized nevi are also frequently located on the trunk (59.2%). Atypical dots/globules (79.6%) and focal hypopigmentation (77.6%) are the most prevalent dermoscopic features in medium-sized nevi. Large-sized nevi are typically located on the trunk (57.1%); atypical dots/globules (100%) and asymmetry (85.7%) are the most common features in large-sized nevi. All sizes of nevi mostly occur in people with Fitzpatrick skin type 4 without any statistical difference.

We designed our study to define distinct dermoscopic features based on size to be helpful in distinguishing CMN from dysplastic nevi and melanoma. The most important conclusions of our study are that in children from our population: The globular pattern as the predominant dermoscopic pattern is more frequent in children younger than 16 years old, and the reticular pattern as the predominant pattern is more frequent in adults older than 16 years old; Atypical dots/globules, focal hypopigmentation, and perifollicular hypopigmentation are the most common dermoscopic features of CMN; The rarest dermoscopic feature is the blue-whitish veil; The presence of atypical dots/globules, asymmetry, and hypertrichosis correlates with the size of CMN; Focal hypopigmentation and milia-like cysts are significantly more common in medium-sized nevi than small-sized and large-sized nevi.

Atypical dots/globules, focal hypopigmentation, perifollicular hypopigmentation, and atypical pigment network have been associated with dysplastic nevi up until now. In recent years, it has been shown that there was a mistake about the terms dysplastic nevus and congenital nevus¹². Most of the small-sized and medium-sized congenital nevi

may have the same dermoscopic patterns and features with dysplastic nevi.

Our findings may provide information for larger studies designed to accurately define the dermoscopic features of CMN.

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