

Re-evaluation of the Anatomical Distribution Patterns in Large Congenital Melanocytic Nevus

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ABSTRACT Introduction: Large congenital melanocytic nevi (LCMNs) are mainly classified according to their distribution patterns, which are thought to reflect their embryological development.

Objectives: We aimed to test the validity of previously described clinical classification systems of LCMNs in a large, well-documented case series.

Methods: The retrospective cohort study including 111 LCMN patients focused on the anatomical distribution of the nevus's main mass.

Results: The trunk was most commonly affected (N=68), followed by the lower extremities (N=35). When the locations of the nevi were evaluated according to the previously described 6B (bolero, back, bathing trunk, breast/belly, body extremity, body patterns) and 7B (6B with the addition of bonce pattern) classification systems, 15 showed the typical bolero pattern, 13 back pattern, 23 bathing trunk pattern, five breast/belly pattern, 25 body extremity pattern, 14 bonce (head) pattern, and one body pattern. Among the 15 patients who could not be fully classified according to these patterns, five were grouped as "hood" (head and neck) pattern, eight as "half-bathing trunk," and two as "atypical bolero" sub-patterns. Isolated lesions on the extremities also exhibited two different sub-patterns: encircling the extremity or not.

Conclusion: Most of our cases demonstrated distribution patterns consistent with the 6B and 7B classification systems. However, the isolated head and neck involvement observed in our series may be described as hood pattern. Furthermore, certain variations, such as those seen in the bathing trunk and breast/belly patterns, may be considered subgroups of these patterns. Isolated extremity lesions of LCMNs exhibited two distinct patterns. These findings suggest that, although existing classification systems provide a valuable framework, further subclassifications may be necessary to account for regional variations observed in clinical practice.

Introduction

Large congenital melanocytic nevi (LCMNs) are rare neural-crest derived hamartomas that can affect almost every part of the body, with varying frequency [1]. In recent years, numerous studies have attempted to classify LCMNs based on their distribution patterns [2-7] and to correlate these patterns with embryological development [8]. However, most of these studies included a limited number of patients and were based solely on retrospective evaluations of electronic medical records from multiple centers or on clinical images published in the literature [2-5,7]. Therefore, the clinical understanding of this subject may still be incomplete.

Objectives

In this single center study, we evaluated a large cohort of LCMN cases to determine the predominant anatomical regions of nevi (main masses) and their characteristic distribution patterns. Our findings were then compared with the existing literature, including current classification systems for nevus distribution [2,3].

Methods

In this retrospective, cross-sectional study, we evaluated 111 consecutive patients diagnosed with LCMN between 2004 and 2020. Nevi located on the trunk or extremities with a projected adult size of ≥ 20 cm or on the head exceeding the size of a patient's own palm were included [2-10]. Although various size classification systems for LCMN exist, these criteria were used to ensure consistency across the study cohort [9-18]. Two types of LCMN were distinguished. The classical type presents as a main mass, usually associated with multiple satellite nevi distributed across the body, while the nevus spilus type consists of variable numbers of small-to-large congenital melanocytic nevi (CMN) superimposed on an extensive (>20 cm) café-au-lait-like macula [19]. Patients who had undergone partial or total excision of the LCMN prior to diagnosis were excluded.

Patient demographics were collected, and participants were stratified into pediatric (<18 years) and adult (≥ 18 years) groups [20]. The body was divided into five main anatomic regions for nevus localization: head, neck, trunk, upper extremities, and lower extremities. These main anatomical regions were further subdivided for detailed evaluation [Table 1]. Anatomical regions affected by the main mass of the nevus (classical LCMN) or café-au-lait-like macula on the base (nevus spilus type LCMN) were determined. The correlation between the regions in the series and well-known LCMN distribution patterns was examined.

IBM SPSS Statistics Version 26 was used to store and analyze the data. Descriptive statistics were calculated as median, minimum, and maximum values for continuous variables and as frequency and percentage for categorical variables. The study was approved by the ethics committee of the university faculty (approval number: 2020/06) and conducted in accordance with the Declaration of Helsinki.

Results

The study included 111 patients (44 males; 39.6%, 67 females; 60.4%), with a mean age at admission of 15.2 + 11.8 years (median: 12, range 0–61 years); 80 were pediatric patients and 31 were adults. Classical LCMN was observed in 106 patients (95.5%), while nevus spilus-type LCMN was seen in five (4.5%) [Figure 1A].

The main mass of the nevus was most commonly located on the trunk (N=68), followed by the lower extremities, upper extremities, head, and neck [Table 1]. While 66 patients (59%) had involvement of a single anatomical region, 45 patients (41%) had nevi affecting multiple regions. The frequency of involvement of each anatomical region by the nevus mass is summarized in Table 1.

The classification of the patients according to the previously defined anatomical distribution patterns of LCMN [2-3] revealed the following frequencies: bathing trunk (N=23) [Figure 1B], body extremity (N=25) [Figure 1C], bonce (N=14) [Figure 1D], back (N=13) [Figure 1E], bolero (N=15) [Figure 1F], breast/belly (N=5) [Figure 2A], and body (N=1) [Table 2]. The remaining 15 patients did not fully conform to previously defined clinical patterns. Ten of these cases were considered as special clinical variants (subgroups) of the bathing trunk and bolero patterns. Nevi involving only the upper part of gluteal region, upper part of the gluteal region and lower back together [Figure 2B], or unilateral iliac region with or without extension to the genital area, gluteal folds, or upper legs [Figure 2C and D] were observed in eight patients and were considered a clinical subgroup of the well-known bathing trunk pattern. The nevi of two patients with anterior trunk involvement extending partially to the back were categorized as atypical bolero pattern due to the absence of neck involvement [Figures 2E and F]. The other five patients had LCMNs limited to the scalp and neck, resembling a distinct pattern called "hood," which has not been previously described as a specific distribution pattern in existing classification systems [Figure 3A].

Among the 25 cases with isolated upper and lower extremity nevi, two previously undescribed distribution patterns were identified: in 11 cases, the nevus encircled the extremity [Figure 1C], whereas in 14 cases, it did not [Figures 3B and C].

Table 1. Frequency of involvement of the anatomical regions by the nevus's main mass in the patients. Abbreviation: LCMN: Large Congenital Melanocytic Nevus.

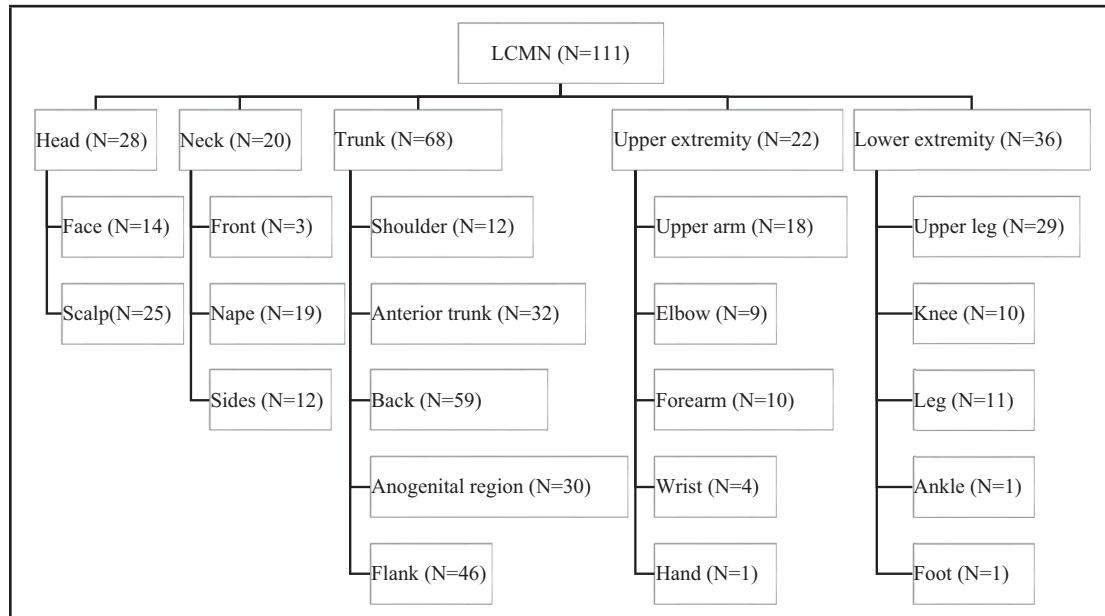


Figure 1. (A) Nevus spilus-like large congenital melanocytic nevus on the lower trunk. (B) Symmetrical large congenital melanocytic nevus on upper legs, genitalia and lower trunk representing classical bathing trunk pattern. (C) Large congenital melanocytic nevus encircling a large part of the arm, representing body extremity pattern. (D) Large congenital melanocytic nevus on the scalp and face. (E) Large congenital melanocytic nevus on the back, representing back pattern. (F) Large congenital melanocytic nevus on the upper back and neck, representing classical bolero pattern.

Among the 28 patients with head involvement, 14 had isolated lesions, and 14 exhibited extension of the main mass to additional body areas. Of the patients with head involvement (regardless of extension to other regions), two presented with isolated facial lesions [Figure 3D], 14 had isolated scalp lesions [Figure 3E], and 12 showed combined scalp and facial involvement [Figures 1D and 3F]. Among

the 14 patients with involvement of other body areas along with the head [Figure 1F], four had both scalp and facial involvement, while 10 had only scalp lesions. Of the 14 patients with LCMN limited to the head region [Figures 3D–F], four had only scalp lesions, two had only facial lesions, and eight had both scalp and facial involvement. Among the 20 patients with neck involvement, five had extension to the

Table 2. Classification of the patients based on the specific location patterns of the nevus' main mass.

Location pattern of nevus' main mass	Case number	Proportion (%)
Breast/belly	5	4.5%
Back	13	11.7%
Bathing trunk	31	27.9%
Classical bathing trunk	23	
Half-bathing trunk*	8	
Bolero	17	15.3%
Classical bolero	15	
Atypical bolero*	2	
Body extremity	25	22.6%
Encircling the extremity*	11	
Not encircling the extremity*	14	
Body	1	0.9%
Head (bonce)*	14	12.6%
Hood*	5	4.5%

*Patterns or subgroups not described in 6B and 7B clinical classifications.



Figure 2. (A) Large congenital melanocytic nevus on the anterior trunk, representing breast/belly pattern. (B) Large congenital melanocytic nevus restricted to bilateral upper gluteal region, classified as half-bathing trunk pattern. (C) Large congenital melanocytic nevus on both sides of the lower back, gluteal area, and right upper thigh, classified as half-bathing trunk pattern. (D) Large congenital melanocytic nevus on located unilaterally iliac region and upper thigh classified as half-bathing trunk pattern. (E, F) Large congenital melanocytic nevus located on anterior trunk, shoulder, and upper back unilaterally but not extending to the neck, which represents atypical bolero pattern.

head, six to the trunk, and nine had involvement of both head and trunk [Figure 1F]. None of the patients had isolated neck lesions.

Of the 19 LCMN patients with lesions confined to the head (N=14) or both the head and neck without extension to neighboring body areas (N=5), the nevi were predominantly distributed unilaterally (N=11) [Figures 3D and F] or along the midline (N=8) [Figure 3E]. Patients with predominantly unilateral involvement were divided into subgroups. The first

subgroup (N=3) had nevi confined to the forehead and scalp. The second subgroup (N=6) presented with lesions extending from the scalp to the nose and eyelids, with partial ear involvement but none extended to the chin. The third subgroup (N=2) also had unilateral head involvement but showed additional extension to the neck. Nevi with midline distribution were likewise classified into three subgroups: those limited to the scalp (N=6) [Figure 3E], those confined to the face (N=1), and those involving both the scalp and face (N=1).



Figure 3. (A) Large congenital melanocytic nevus restricted to the scalp and neck, defined as hood pattern. (B) Large congenital melanocytic nevus located only on one side of the thigh. (C) Large congenital melanocytic nevus on the pretibial area which is not circumferential to the extremity, resembling a shin guard. (D) Large congenital melanocytic nevus on the nose, periorbital area, and forehead showing unilateral distribution. (E) Large congenital melanocytic nevus on the midline of the scalp. (F) Large congenital melanocytic nevus limited to scalp and face in unilateral distribution.

Discussion

Our study demonstrated that the main distribution patterns of the nevi were generally consistent with the classical 6B and 7B classification systems, which include terms such as breast/belly, back, bathing trunk, bolero, body extremity, body, and bonce [2,3]. In addition, we identified a novel distribution pattern and distinct variants of known patterns that may constitute their subgroups.

LCMNs can involve a single anatomical region (59% in our series) or multiple regions (41% in our series), with patterns often named anatomically for single-region involvement (e.g., back, breast/belly, body extremity, bonce) and after clothing items for multi-region involvement (e.g., bolero, bathing trunk) [2,3,7]. The “body” pattern denotes involvement of multiple anatomical regions. The 6B rule was originally defined for giant CMN (projected adult size >40 cm), while the 7B rule, which additionally includes “bonce” (head) was proposed for LCMN cases (>20 cm) [3]. Our series, consisting of 93 LCMNs with projected adult size over 20 cm (excluding the head) and 19 lesions located on the head and/or neck larger than one palm area of the patient’s own hand was evaluated within the framework of these classification systems.

In a study by Kinsler and Larue, which examined the distribution of pigmented lesions in animal models to explore their embryological origins, CMNs were found to develop in a non-segmental pattern [8]. Thirteen distinct distribution areas were identified, including three body regions

(two dorsal, one ventral), six extremity regions (four distal, two middle), and four head regions (one dorsal, three ventral), each corresponding to a specific melanocyte precursor [8]. The ventral body area described by Kinsler and Larue [8] clinically corresponds to the “breast/belly” pattern, while the upper dorsal region, extending from the cranial vertex on through the neck and arms and down to the lumbar area, matches the classical “bolero” pattern in existing classifications [2,3]. In addition to the 15 cases exhibiting the typical bolero pattern in our series, we identified two LCMNs that were primarily located within the same anatomical region but partially differed in distribution. These lesions mainly located within this specific area did not extend to the neck; instead, they involved the anterior trunk, spreading from flank to back [Figures 2E and F]; although they partially resembled the breast/belly pattern due to anterior trunk involvement, they were not confined to that region. Therefore, we called them “atypical bolero” pattern. The lower dorsal body area, starting from the upper thoracic vertebrae and covering the gluteal region and perineum, extending to the upper part of the legs [8], corresponds to the “bathing trunk” pattern described in clinical classifications [2,3]. In our series, 23 cases displayed this pattern. However, eight LCMNs did not encircle the lower body like a typical bathing trunk and did not extend to both legs; instead, they were usually restricted to the upper gluteal region and lower back with limited extensions [Figures 2B and C] or were unilaterally located in the same region [Figure 2 D]. We propose describing these as “half-bathing trunk” pattern.

According to the description by Kinsler and Larue, a total of four specific areas are defined in lower extremities: two distal region (left and right), extending from the toes to just below the knee (stocking-type), and two middle regions (left and right), extending from the mid-thigh to the mid-calf [8]. In contrast, only two specific areas have been identified in the upper extremities—one on each side—extending from the fingertips to the shoulders [8]. The “body extremity” pattern used in clinical classifications aligns with the extremity regions defined in embryological studies [2,3]. In our cohort, analysis of involvement frequencies across these regions revealed a predominance of proximal localization over distal localization in both the upper and lower limb [Table 1]. This finding was interpreted as reflecting the involvement of the proximal extremities as a part of the dorsal body area. Notably, we observed LCMNs in the upper extremities that resembled the anatomical distribution patterns previously described as the middle extremity areas of the lower limbs. This observation raises the possibility that a middle segment may also exist in the upper extremities. Additionally, we identified two previously undescribed distribution patterns in patients with isolated nevi of the upper and lower extremities: one characterized by circumferential (encircling) extremity-like involvement resembling a knee pad and bracelet (N=11) [Figure 1C] and the other by lesions confined to one side of limb, resembling a shin guard (N=14) [Figure 3C]. Therefore, nevi with encircling involvement were aligned with the distal extremity areas, while those with partial involvement were more consistent with the middle extremity areas described by Kinsler and Larue. Further studies should evaluate whether these newly defined extremity subgroups possibly characterized by distinct embryological origins hold clinical prognostic significance. Their standardized classification could provide clinicians with a practical framework for describing complex nevus patterns that would otherwise require overly detailed topographic descriptions.

In our study, head involvement was observed in 28 patients, with isolated lesions occurring in half of the cases (N=14). Among the remaining patients, this presentation was associated with the bolero pattern in eight cases and with the body pattern in one case. The other five patients exhibited a distinct distribution extending from the vertex to the neck [Figure 3A], a pattern not previously defined in LCMN classifications, we propose the term “hood” to describe this scalp-neck continuum. For the 14 isolated head cases, lesions were further categorized as scalp only, face only, or combined scalp-facial involvement. Given this anatomical specificity, “head” (rather than the 7B rule “bonce”) better reflects the observed patterns. Incorporating these observations including the addition of “hood” pattern and the replacement of the term “bonce” with “head” [Table 2], we suggest to update the terminology of anatomical distribution in LCMN classification as “6B+2H rule”.

While the 7B classification acknowledges the head-region involvement, it does not define specific clinical patterns of these nevi [3]. Embryologically, the head is divided into dorsal and ventral cranial areas [8]. The dorsal area demonstrates bilateral symmetry along the midline, particularly near the vertex, whereas the ventral area comprises three midline-centric regions (upper, middle, and lower face). In our cohort, head nevi clustered into two groups: lesions perfectly matching these predefined embryological zones and nevi involving multiple contiguous zones simultaneously. This heterogeneity underscores the need for larger-scale studies to establish a standardized pattern classification for cranial lesions.

Clinical pattern classifications generally align with embryologically defined areas. They essentially map the territorial expansion of melanocyte precursor cells, which, depending on the timing of postzygotic mutations—a key etiological factor—across a broad spectrum. This spectrum ranges from extensive LCMNs encompassing entire anatomical regions to a small CMNs arising from a single melanocyte clone [20,21]. Notably, we encountered limited forms of classic patterns (bathing trunk, bolero, and body extremity variants), all of which can be mechanistically interpreted through this developmental framework.

Conclusion

For LCMNs exhibiting isolated head and neck involvement, we propose the term “hood” distribution pattern. Lesions predominantly localized on the bathing trunk or bolero regions but demonstrating limited anatomical extension may be classified as “half-bathing trunk” and “atypical bolero” variants, respectively. LCMNs limited to the extremities manifest two distinct patterns: encircling involvement, resembling a knee pad or bracelet configuration, and non-encircling lesions, mimicking a shin guard distribution. While the classical 6B and 7B classification systems cover most cases, our observations suggest that terminology revisions are necessary to account for these pattern variations.

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