Patterns of distribution of giant congenital melanocytic nevi (GCMN): The 6B rule

Vanessa P. Martins da Silva, MD,[a] Ashfaq Marghoob, MD,[b] Ramon Pigem, MD,[a] Cristina Carrera, PhD,[a,c] Paula Aguilera, PhD,[a] Joan A. Puig-Butillé, PhD,[c,d] Susana Puig, PhD,[a,c] and Josep Malvehy, PhD[a,c]
Barcelona, Spain, and New York, New York

Background: Garment-related terms have been used to describe the pattern of distribution of giant congenital melanocytic nevi (GCMN).

Objective: We sought to describe patterns of distribution of GCMN and propose a classification scheme.

Methods: Photographic records of patients with GCMN from the Hospital Clinic of Barcelona were analyzed and a classification based on observed GCMN distribution patterns was created. The classification was independently applied by 8 observers to cases found in the literature. The interobserver agreement was assessed.

Results: Among 22 patients we observed 6 repeatable patterns of distribution of GCMN, which we termed the "6B": bolero (involving the upper aspect of the back, including the neck), back (on the back, without involvement of the buttocks or shoulders), bathing trunk (involving the genital region and buttocks), breast/belly (isolated to the chest or abdomen without involvement of bolero or bathing trunk distributions), body extremity (isolated to extremity), and body (both bolero and bathing trunk involvement). Our literature search found 113 cases of GCMN, which we were able to classify into 1 of the 6B patterns with an overall kappa of 0.89.

Limitations: Some patterns occur infrequently with a dearth of images available for analysis.

Conclusions: The anatomic distribution of GCMN occurs in 6 recognizable and repeatable patterns. (J Am Acad Dermatol 2017;76:689-94.)

Key words: benign skin tumors; classification; congenital melanocytic nevus; distribution; giant nevus; skin neoplasms.

From the Melanoma Unit, Department of Dermatology, Hospital Clinic of Barcelona, University of Barcelona[a]; Department of Dermatology, Memorial Sloan Kettering Cancer Center, New York[b]; "Centro de Investigación Biomédica en Red en Enfermedades Raras, Instituto de Salud Carlos III," Barcelona[c]; and Department of Biochemical and Molecular Genetics, Hospital Clínic, IDIBAPS, University of Barcelona.[d]

The research at the Melanoma Unit of the Hospital Clinic of Barcelona is partially funded by grants 03/0019, 05/0302, 06/0265, 09/01393, and 12/00840 from Fondo de Investigaciones Sanitarias, Spain; by the Centro de Investigación Biomédica en Red de Enfermedades Raras, Instituto de Salud Carlos III, Spain; by the AGAUR 2009 SGR 1337 and AGAUR 2014 SGR 603 of the Catalan Government, Spain; by the European Commission under the Sixth Framework Program, contract number LSHC-CT-2006-018702 (GenoMEL), and under the Seventh Framework Program (Diagnoptics); by the National Cancer Institute of the US National Institutes of Health (CA83115); by the MARATÓ de TV3 Foundation; by the Telemaraton Foundation (research grant "Todos somos raros"); and by the Leo Messi Foundation. The project "Phenotype and molecular characterization of large to giant congenital melanocytic nevi" has received funding from the Spanish Federation of Neuromuscular Disease, the Spanish Federation of Rare Diseases, and Isabel Gemio Research Foundation for muscular dystrophy and other rare diseases through the call for research projects on rare diseases—2014 through the initiative "We are rare, all are unique." The funders had no role in the design and conduct of the study; in the collection, analysis, and interpretation of the data; in the decision of the submission of the manuscript; or in the preparation, review, or approval of the manuscript.

Conflicts of interest: None declared.

Accepted for publication May 30, 2016.

Reprint requests: Josep Malvehy, PhD, Melanoma Unit, Department of Dermatology, Hospital Clinic of Barcelona, C/Villarroel, 170. Esc 3-4, 08036 Barcelona, Spain. E-mail: jmalvehy@gmail.com.

0190-9622

© 2016 by the American Academy of Dermatology, Inc. Published by Elsevier, Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

http://dx.doi.org/10.1016/j.jaad.2016.05.042

Congenital melanocytic nevi (CMN) have an estimated prevalence of between 0.5% and 31.7%.[1-5] CMN larger than 20 cm are uncommon with an estimated incidence of between 1 in 20,000 to 1 in 500,000 births.[2,6] The current classification of CMN (Fig 1) is based mainly on the maximum diameter that the nevus is predicted to attain during adulthood.[7] A CMN is classified as large-1 if it measures between 20 to 30 cm, large-2 if it measures between 30 and 40 cm, giant-1 if it measures between 40 and 60 cm, and giant-2 if greater than 60 cm in maximal diameter.[7] This classification scheme also includes additional morphologic characteristics of the CMN: anatomic localization, degree of color heterogeneity, surface rugosity, hypertrichosis, dermal/subcutaneous nodularity, and number of smaller satellite nevi.

CAPSULE SUMMARY

- Garment-related terms have been used to describe the anatomic localization of giant congenital melanocytic nevi.
- A classification scheme, with good interobserver agreement, is proposed to standardize the distribution patterns of giant melanocytic nevi.
- A simple, reliable, and repeatable classification can help to improve communication and patient risk stratification.

To highlight the distribution of giant CMN (GCMN), Bircher,[8] in 1897, used terms such as "bathing trunk," "vest-like," and "vest with collar." In 1965, Reed et al[9] described 55 patients using terms such as "bathing trunk," "shoulder stole" or "cape-like," "coat sleeve," and "stocking-like." In 2005, Torrelo et al[10] separated 1188 nevi based on the following morphologic categories: round, patchy indented or triangular shape, agminated, diffuse patchy, Blaschkolinear, block/flag-like, and garment-like distribution.

Based on the garment-related terms used to describe the distribution patterns of large or GCMN, we hypothesized that GCMN have repetitive recognizable patterns of distribution. The aim of this study was to evaluate the distribution patterns of GCMN and to determine whether recognizable patterns emerge.

METHODS

Phase I

A query of the medical records at the Hospital Clinic of Barcelona was performed to identify patients diagnosed with GCMN between 1975 and 2013. The charts were reviewed to identify patients who had full-body clinical photographs of CMN with a projected adult size larger than 40 cm and in whom the full extent (location, distribution, and size) of the GCMN was visible on the photographs. The included cases were analyzed by 2 observers to determine the patterns of distribution and to create an anatomic distribution classification with initial definitions and schematics termed as the "6B" (Fig 2). Two other observers independently reclassified the same cases using the proposed classification scheme. Cases that presented disagreement in the classification were jointly re-evaluated for consensus. This exercise lead to fine-tuning of the classification and the final schematic was created (Fig 2). Two different observers independently classified the cases according to the proposed final classification scheme and interobserver agreement was assessed.

Phase II

To test the reproducibility and applicability of the 6B classification scheme, an atlas of published images of patients with GCMN was assembled. All articles identified via literature search using PubMed containing the terms "giant congenital nevi" and published between January 1, 1998, and April 30, 2014, were evaluated for the presence of images of GCMN. Images were selected if they met the following criteria: (1) images showing the GCMN with a projected adult size larger than 40 cm in which the entire GCMN was visible with back, front, and/or side body sectors shown; and (2) images showing the main portion of the GCMN with a written description that adequately helped the reader appreciate the full extent of its distribution. Images excluded from the study atlas included those showing only a portion of the GCMN where the full extension of the GCMN could not be deciphered, case reports that were deemed not to represent CMN or GCMN, duplicate images of the same patient in multiple publications, and GCMN where portions of the nevus had been excised.

The workflow for phase II of this study is shown in Fig 3. All cases included in the study atlas were classified according to the 6B classification scheme by 8 physicians. Two weeks later, 4 of the 8 physicians classified all cases for a second time. They did not have access to the answers previously provided. The new answers were used to assess the intraobserver agreement.

Statistical analysis

All statistical analyses were performed using software (SPSS Statistics for Windows, Version 22.0, IBM Corp, Armonk, NY). Descriptive data were

Download English Version:

https://daneshyari.com/en/article/5648129

Download Persian Version:

https://daneshyari.com/article/5648129

Daneshyari.com