The scalp hair collar and tuft signs: A retrospective multicenter study of 78 patients with a systematic review of the literature

Didier Bessis, MD,[a,y] Michèle Bigorre, MD,[b] Nausicaa Malissen, MD,[a] Guillaume Captier, MD, PhD,[b] Christine Chiaverini, MD,[c] Claire Abasq, MD,[d] Sébastien Barbarot, MD, PhD,[e] Olivia Boccara, MD,[f] Emmanuelle Bourrat, MD,[g] Hassan El Fertit, MD,[h] Catherine Eschard, MD,[i] Thomas Hubiche, MD,[j] Jean-Philippe Lacour, MD, PhD,[c] Nicolas Leboucq, MD,[k] Emmanuel Mahé, MD, PhD,[l] Stéphanie Mallet, MD,[m] Myriam Marque, MD,[n] Ludovic Martin, MD, PhD,[o] Juliette Mazereeuw-Hautier, MD, PhD,[p] Nathalie Milla, MD,[a] Alice Phan, MD, PhD,[q] Patrice Plantin, MD,[r] Marie-Christine Picot, MD, PhD,[s] Eve Puzenat, MD,[t] Valérie Rigau, MD, PhD,[u] Pierre Vabres, MD, PhD,[v] Sylvie Fraitag, MD,[w] and Franck Boralevi, MD, PhD,[x] on behalf of the Groupe de Recherche Clinique en Dermatologie Pédiatrique

Montpellier, Nice, Brest, Nantes, Paris, Nîmes, Reims, Fréjus, Argenteuil, Marseille, Angers, Toulouse, Lyon, Quimper, Besançon, Dijon, and Bordeaux, France

Background: Hair collar sign (HCS) and hair tuft of the scalp (HTS) are cutaneous signs of an underlying neuroectodermal defect, but most available data are based on case reports.

Objective: We sought to define the clinical spectrum of HCS and HTS, clarify the risk for underlying neurovascular anomalies, and provide imaging recommendations.

Methods: A 10-year multicenter retrospective and prospective analysis of clinical, radiologic, and histopathologic features of HCS and HTS in pediatric patients was performed.

Results: Of the 78 patients included in the study, 56 underwent cranial and brain imaging. Twenty-three of the 56 patients (41%) had abnormal findings, including the following: (1) cranial/bone defect (30.4%), with direct communication with the central nervous system in 28.6%; (2) venous malformations (25%); or (3) central nervous system abnormalities (12.5%). Meningeal heterotopia in 34.6% (9/26) was the most common neuroectodermal association. Sinus pericranii, paraganglioma, and combined nevus were also identified.

Limitations: The partial retrospective design and predominant recruitment from the dermatology department are limitations of this study.

From the Departments of Dermatology at Saint-Eloi Hospital,[a] Montpellier, Archet-2 Hospital,[c] Nice, Brest University Hospital,[d] Brest, Hôtel-Dieu Hospital,[e] Nantes, Robert-Debré Hospital,[i] Reims, Argenteuil Hospital,[l] Argenteuil, Timone Hospital,[m] Marseille, Caremeau Hospital,[n] Nîmes, Angers University Hospital,[o] Angers, National Center for Rare Skin Disorders,[p] Larrey Hospital, Toulouse, Quimper Hospital,[r] Quimper, Besançon University Hospital,[t] Besançon, and Bocage Hospital and Bourgogne Medical University,[v] Dijon; Department of Infantile Plastic Surgery,[b] Lapeyronie Hospital, Montpellier; Departments of Pediatric Dermatology at Necker-Enfants Malades Hospital[f] and Robert-Debré Hospital,[g] Paris, Femme-Mère-Enfant Hospital and Claude-Bernard Lyon 1 University,[q] Lyonm and the National Center for Rare Skin Disorders-Institut National de la Santé et de la Recherche Médicale (INSERM) U1035[x]; Department of Infantile Neurosurgery,[h] Caremeau Hospital, Nîmes; Department of Dermatology and Infectious Diseases,[j] Fréjus Hospital, Fréjus; Department of Neuroradiology,[k] Gui de Chauliac Hospital, Montpellier; Unit of Clinical Research and Epidemiology,[s] Department of Medical Information, Antonin Balmes Hospital, Montpellier; Department of Pathology,[u] Montpellier University Hospital, Montpellier; Department of Pathology,[w] Necker-Enfants Malades University Hospital, Paris; and Montpellier University Hospital[y] and Institut National de la Santé et de la Recherche Médicale (INSERM) U1058, Montpellier.

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Reprint requests: Didier Bessis, MD, Department of Dermatology, Saint-Eloi Hospital, CHRU Montpellier, 80, Ave Augustin Fliche, 34295 Montpellier Cedex, France. E-mail: d-bessis@chu-montpellier.fr.

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Conclusions: Infants with HCS or HTS are at high risk for underlying neurovascular anomalies. Magnetic resonance imaging scans should be performed in order to refer the infant to the appropriate specialist for management. (J Am Acad Dermatol 2017;76:478-87.)

Key words: cephalocele; hair collar; hair tuft; membranous aplasia cutis; meningocele; paraganglioma; sebaceous nevus; sinus pericranii; smooth muscle cell nevus.

INTRODUCTION

The hair collar sign (HCS) was reported in 1989 by Commens et al[1] to describe a congenital collar of hair surrounding a bald scalp lesion. It was found to be a marker for neuroectodermal defects, such as heterotopic brain tissue or cephalocele.[2] Subsequently, HCS was described overlying other congenital lesions, such as nevus sebaceus[3] or arteriovenous fistula[4] and as part of the spectrum of oculoectodermal syndrome,[5] without evidence of a neuroectodermal defect. Rare observations of other scalp hair abnormalities, including congenital midline hair tuft of the scalp (HTS) or small tufts of hair extruding from an orifice, were reported to overlie atretic meningocele or meningeal heterotopia.[6,7] Data on these congenital scalp hair abnormalities have been drawn mostly from limited case reports, with a single retrospective series of 10 observations.[8] We investigated the clinical characteristics, cranial and central nervous system (CNS) imaging findings, and histopathologic findings in 78 new cases of HCS and/or HTS. We reviewed 66 previously published cases to define the clinical spectrum, clarify the risk for underlying neuroectodermal and cerebrovascular anomalies, and provide imaging recommendations.

CAPSULE SUMMARY

- The hair collar sign and hair tuft of the scalp are signs of a potential underlying neuroectodermal defect.
- The cutaneous findings are not predictive of a specific cranial or central nervous system anomaly.
- Magnetic resonance imaging should be performed routinely in these patients.

METHODS

This retrospective and subsequently prospective study was conducted in 20 French Departments of Dermatology (ie, Angers, Argenteuil, Besançon, Brest, Dijon, Fréjus, Lyon, Marseille, Montpellier, Nantes, Nîmes, Nice, Quimper, Reims, and Toulouse), Pediatric Dermatology (ie, Bordeaux and Paris [Robert Debré and Necker Hospitals]), Infantile Plastic Surgery (Montpellier), Pediatric Neurosurgery (Nîmes), and by the Groupe de Recherche Clinique en Dermatologie Pédiatrique.

All patients <18 years of age with HCS or HTS seen by pediatric dermatologists were retrospectively included from 2005 to 2009 and prospectively included from 2010 to 2014. HCS was defined as a ring of long, thick, coarse hair surrounding a nodule, cyst, or bullous and/or atrophic hairless area on the scalp. HTS was defined as a prominent tuft of long, thick, coarse hair on the scalp without an apparent central lesion. The age, sex, associated conditions, and physical features, including number, location, size, and morphologic appearance of HCS and HTS were documented in all patients. In the absence of a clear consensus on the use of the term "vertex" among investigators and imprecision or variability in the literature,[9] we defined the vertex as a median circular area extending from the posterior edge of the anterior fontanel to the posterior edge of the posterior fontanel for simplicity and reproducibility. The "membranous" variant of aplasia cutis (MAC) was defined by the presence of a thin, glistening, parchment-like membranous epithelial covering at the surface or filled with serous fluid, giving the appearance of a blister.[10] We used a classification of cranial congenital neural tube defects that appeared appropriate for dermatologic, neurosurgical, and pathologic findings.[2,10] Therefore, meningocele was defined as a skin-covered lesion containing meningothelial elements and cephalocele as containing neuroglial tissue with or without meningothelial tissue—these 2 variants corresponding to direct extension into the skin from an intracranial lesion (ie, "classical" or "communicating"). Meningeal heterotopia (ie, "rudimentary meningocele," "meningeal hamartoma," "meningothelial hamartoma," "cutaneous meningioma," or "sequestrated meningocele") or neuroglial heterotopia were defined by the presence, respectively, of meningeal or neuroglial tissue in the absence of a connection, even minor, to the CNS or underlying bone defects noted on radiologic or perioperative data. The terms "atretic meningocele" or "atretic cephalocele" were used only when a rudimentary connection was present and identified during surgery, such as a fibrous tract to bone or dura. Clinical