

Dermoscopy of Cutaneous Neurocristic Hamartoma and Report of its Rare Clinical Presentation

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Keywords: dermoscopy, cuticis verticis gyrata, neurocristic hamartoma, Blue Naevi

Citation: Peter D, Kuryan P, Gupta AK, Thomas M. Dermoscopy of cutaneous neurocristic hamartoma and report of its rare clinical presentation. *Dermatol Pract Concept*. 2022;12(3):e2022138. DOI: <https://doi.org/10.5826/dpc.1203a138>

Accepted: December 27, 2021; **Published:** July 2022

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Funding: None.

Competing interests: None.

Authorship: All authors have contributed significantly to this publication.

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Introduction

Cutaneous neurocristic hamartoma (NCH) is a rare hamartomatous proliferation of melanocytic, neuroid, and mesenchymal tissues first described by Tuthill et al in 1982 [1].

Case Presentation

A 33-year-old man presented with swelling of the scalp and multiple bluish swellings on the body since birth. He had a surgical excision of a scalp lesion at four years of age after which the swelling had recurred. There was a gradual increase in size of the swelling since the past four years. On examination, he had a cerebriform swelling with alopecia, akin to cuticis verticis gyrata with areas of bluish color and depigmentation seen over the occipital scalp and extending onto the neck. (Figure 1A). Multiple bluish firm nodules were seen over the trunk (Figure 1B). The skin lesions on dermoscopy (FotoFinder Systems GmbH) showed varying

colors from blue, gray, white and brown color. The dermoscopy of nodules showed blue clods over a gray background with peripheral brown pigment network (Figure 1C), blue gray clods on a white background (Figure 1D) blue clods on a white background with brown dots (Figure 1E). The scalp nodule showed blue gray clods, white featureless areas, brown dots and streaks, multiple clustered dotted and linear vessels (Figure 2A). The clinical differential diagnoses considered were blue nevus, cerebriform intradermal nevi, nevus sebaceous and cylindroma. An excision biopsy and split thickness skin grafting of the scalp lesion was done. Histopathology from the scalp and trunk nodule showed proliferated spindle to epithelioid nevus cells in dermis and intervening stroma showing a proliferation of bland spindle shaped cells, few of which had wavy nuclear contour (Figure 2, B and C). On immunohistochemistry, the nevus cells were positive for S100, HMB 45 and Melan A and stromal spindle cells for CD 34 (Figure 2, D and E). The MIB index was less than 1%. A diagnosis of benign NCH was made.

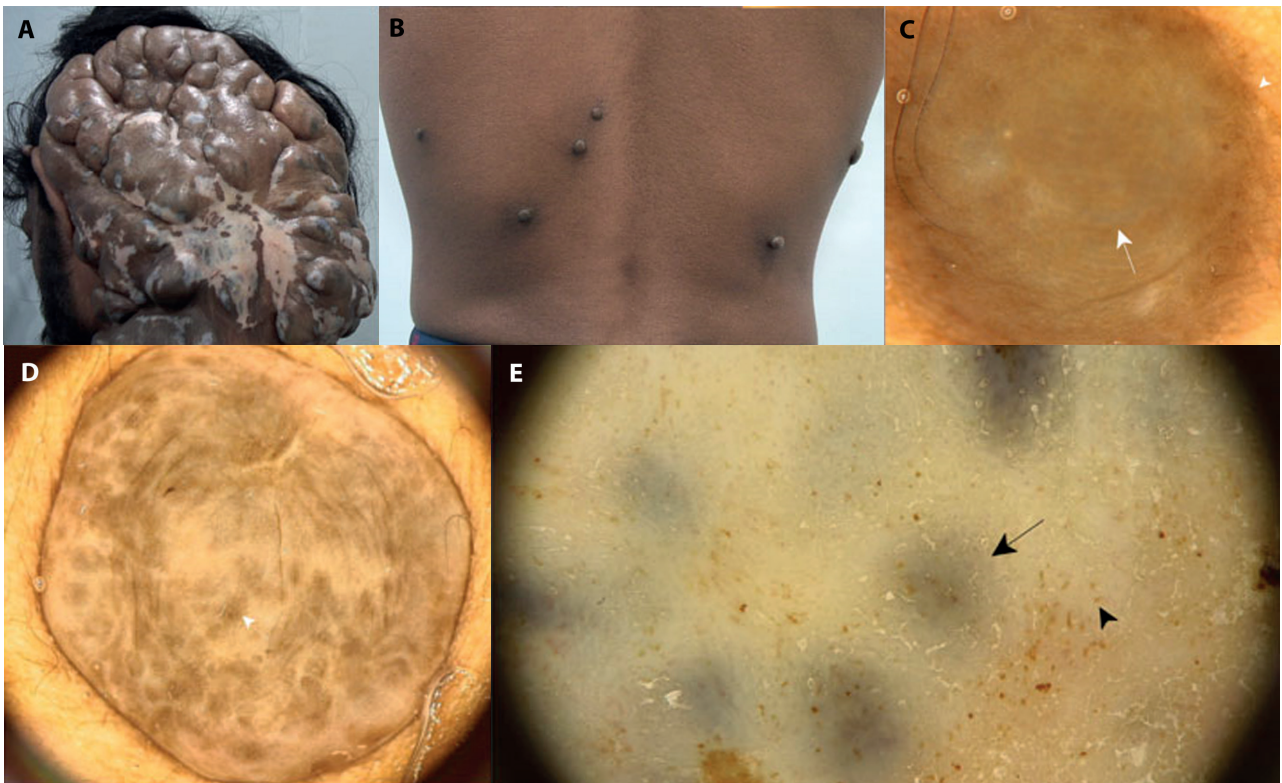


Figure 1. (A) Cerebriform swelling with alopecia, akin to cuticis verticis gyrata with areas of bluish color and depigmentation seen over the occipital scalp. (B) Multiple bluish firm nodules over the back. (C) Dermoscopy of skin nodule showing blue clods (white arrow) over a gray background with peripheral brown pigment network (white arrow head, (20x, non-polarized, fotofinder GmBH). (D) Dermoscopy of skin nodule showing blue gray clods (white arrow) on a white back ground (20X, non-polarized, fotofinder, GmBH). (E) Dermoscopy of nodule over soles showing blue clods (black arrow) on a white background with brown dots (black arrowhead, (20x, non-polarized, fotofinder, GmBH).

Cutaneous neurocristic hamartoma is a rare hamartomatous proliferation of melanocytic, neuroid, and mesenchymal tissues. They show variable differentiation including melanocytic (nevroid, spindle cell, and dendritic), neurosustentacular (Schwann cell and perineural cells), and mesenchymal fibrogenic elements [2]. Congenital or acquired lesions present as pigmented macules, papules or nodules with or without alopecia. There is a predilection for the scalp. Benign NCH presenting as cutis verticis gyrata is very rare but has been reported [3].

Neurocristic hamartoma is a dermal dendritic melanocytic proliferation under borderline category [4] and shares histological features with blue and congenital nevi. Clinically,

histopathologically, and even dermoscopically NCH can resemble a blue nevus. Dermoscopy of blue nevi showed a homogeneous, structureless pigment pattern, with either blue, white–blue, black, brown, and polychromatic [4]. The white–blue color was attributed to the oval/spindle cells the deeper location of the lesions within the dermis and the presence of a thicker subepidermal grenz zone and was seen more with hypochromic blue nevi [4]. Our patient had polychromatic lesions but predominantly blue gray and white clods. On histopathology had epithelioid/spindle cells with deep reticular dermis extension. The course of NCH is generally indolent, however, longstanding NCH has been reported to give rise to cutaneous malignant melanocytic neurocristic tumors [5].

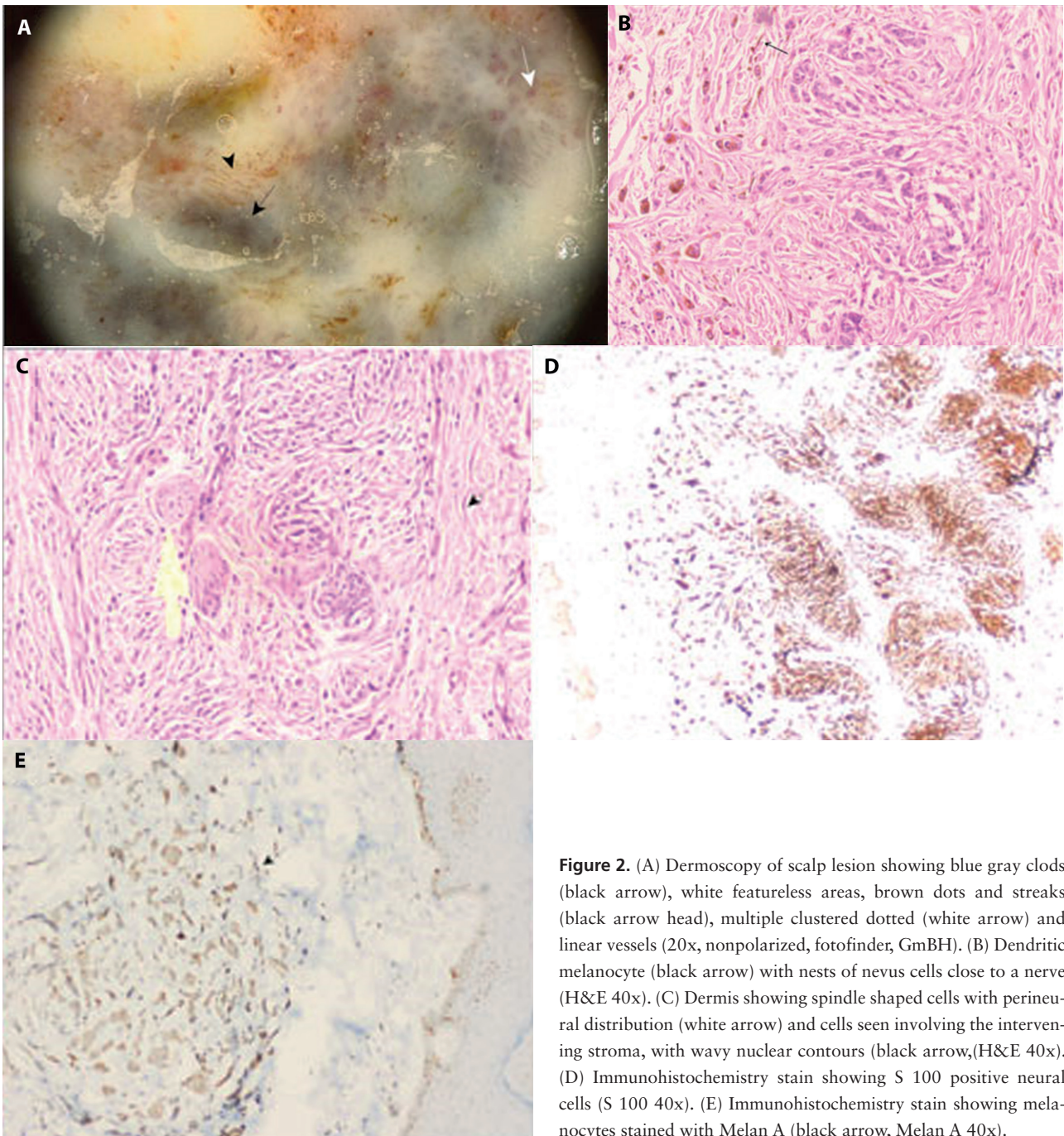


Figure 2. (A) Dermoscopy of scalp lesion showing blue gray clods (black arrow), white featureless areas, brown dots and streaks (black arrow head), multiple clustered dotted (white arrow) and linear vessels (20x, nonpolarized, fofinder, GmbH). (B) Dendritic melanocyte (black arrow) with nests of nevus cells close to a nerve (H&E 40x). (C) Dermis showing spindle shaped cells with perineural distribution (white arrow) and cells seen involving the intervening stroma, with wavy nuclear contours (black arrow, H&E 40x). (D) Immunohistochemistry stain showing S 100 positive neural cells (S 100 40x). (E) Immunohistochemistry stain showing melanocytes stained with Melan A (black arrow, Melan A 40x).

Conclusions

Cutaneous NCH presenting as cutis verticis gyrata is rare. We have described the dermoscopic findings seen in our patient.

References

1. Tuthill RJ, Clark WH, Levene J. Pilar neurocristic hamartoma: its relationship to blue nevus and equine melanotic disease. *Arch Dermatol.* 1982;118(8):592–596. DOI: 10.1001/archderm.118.8.592. PMID: 7103529.
2. McKee PH, Calonje E, Granter SR. Melanocytic nevi. In: McKee PH, Calonje E, Granter SR, eds. *Pathology of the Skin With Clinical Correlations.* 2nd ed. Philadelphia, PA: Elsevier Mosby; 2005:1241–1308.
3. Saha D, Kini UA, Kini H. Cutaneous Neurocristic Hamartoma Presenting as Cutis Verticis Gyrata. *Am J Dermatopathol.* 2014; 36(3):e66–69. DOI: 10.1097/DAD.0b013e3182919c76. PMID: 23812020.
4. Ferrara G, Soyer HP, Malvey J, et al. The many faces of blue nevus. A clinicopathologic study. *J Cutan Pathol.* 2007;34(7): 543– 551. DOI: 10.1111/j.1600-0560.2006.00650.x. PMID: 17576333.
5. Linskey KR, Dias-Santagata D, Nazarian RM, et al. Malignant neurocristic hamartoma: a tumor distinct from conventional melanoma and malignant blue nevus. *Am J Surg Pathol.* 2011; 35(10):1570-1577. DOI: 10.1097/PAS.0b013e31822389b7. PMID: 21934481.