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# Cerebriform intradermal nevus presenting as cutis verticis gyrata: A rarity

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## ABSTRACT

Cutis verticis gyrata (CVG) is a rare skin disorder characterized by cerebriform appearance of the scalp with convoluted folds and deep furrows. Cerebriform intradermal nevus (CIDN) is one of the rare causes of CVG. We report a case of CIDN presenting as CVG on the left parietal area of scalp since birth in a 15-year-old girl. She had no neurological or ophthalmologic manifestations or any other cutaneous lesions.

**Keywords:** Cerebriform intradermal nevus, Computed tomogram, Cutis verticis gyrata, Histopathology, Rare presentation

## INTRODUCTION

Cutis verticis gyrata (CVG) also known as cutis verticis plicata, paquidermia verticis gyrata, and "bulldog" scalp syndrome, is a rare cutaneous disorder characterized by cerebriform appearance of the scalp skin with convoluted folds and deep furrows.[1,2] It may affect the whole scalp or may be localized to one area.

It was originally described by Jean-Louis-Marc Alibert in 1837 and the term CVG was proposed by Unna in 1907. In 1953, Polan and Butterworth classified CVG into primary and secondary.[1] The primary is subdivided into primary essential without any comorbidities,[1] and non-essential form with ophthalmological and neurological manifestations.[1,3] The secondary form is associated with cerebriform intradermal nevus, pituitary tumors, pachydermoperiostosis,[1,4] and various syndromes.[5,6]

## CASE REPORT

A 15-year-old girl with good scholastic performance presented with thickening and pigmentation of the left half of scalp since birth. The lesion started as a pigmented alopecic flat lesion at birth. It started increasing in size and became a raised lesion by 5 years and gradually reached the present size. There was no family history of similar lesions. She had no history of seizures or visual disturbances. Dermatological examination revealed a well-defined pigmented, cerebriform plaque in the left parietal area of size 13 × 12 cm with convolutions on the surface and sparse hair [Figure 1]. She had no other cutaneous lesions. Ophthalmologic and neurologic evaluations and routine blood tests were normal.

Histopathology showed mild hyperkeratosis, and a well-circumscribed lesion involving the upper and mid dermis composed of closely arranged small nests of nevus cells with moderate amount of cytoplasm and melanin without atypia, increased mitosis or junctional activity, suggestive of intradermal nevus [Figure 2].

Ultrasound examination of the scalp showed thickened skin and subcutaneous tissue with absence of color uptake, thus ruling out any underlying vascular malformation [Figure 3a]. Contrast CT (computed tomography) brain with 3D surface rendering showed a non-enhancing circumscribed wavy scalp thickening with tiny specks of calcifications involving the left parietal region with normal calvarium and brain parenchyma [Figures 3b and c]. Thus, a final diagnosis of CIDN presenting as cutis verticis gyrata was made.

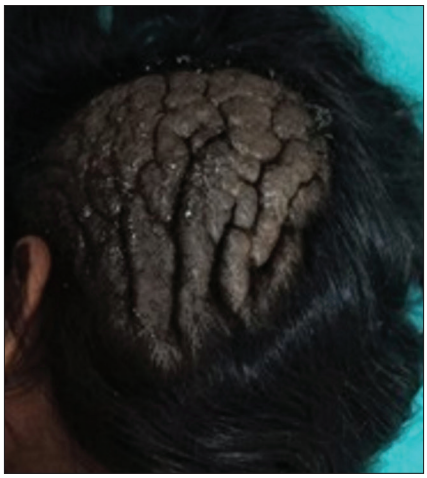

**Figure 1:** A well-defined pigmented, cerebriform plaque of cerebriform intradermal nevus in the left parietal area of size 13 × 12 cm with sparse hair and convolutions on the surface.

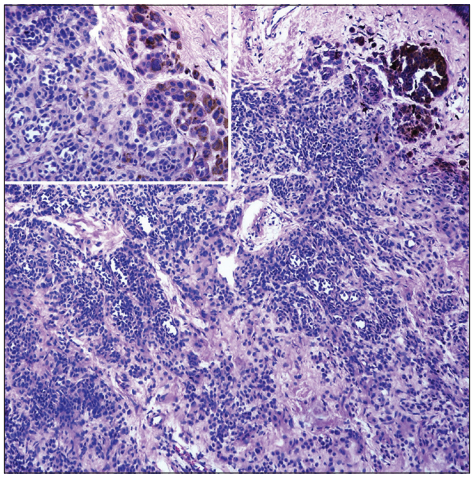

**Figure 2:** Skin biopsy from cerebriform intradermal nevus showing a well-circumscribed lesion involving the upper and mid dermis composed of closely arranged small nests of nevus cells with moderate amount of cytoplasm and melanin without atypia, increased mitosis or junctional activity (H and E, 100×); inset: Higher magnification of the same specimen (H and E, 400×).

## DISCUSSION

Cutis verticis gyrata is a very rare condition, the prevalence being 1 in 100,000.[7] Cerebriform intradermal nevus (CIDN) is a rare cutaneous disorder that can cause secondary CVG. Clinically, CIDN is characterized by a well-demarcated cerebriform plaque, usually on the parietal area of the scalp.[8,9]

The first case of CVG secondary to CIDN on the scalp was reported by Hammon and Ransom in 1937.[9] A case of CIDN, similar to our case, characterized by a pigmented plaque with alopecia on the left parietal and temporal areas of the scalp with typical histology showing solitary and clusters of nevus cells in the dermis has been reported by Alcántara González *et al*.[8] Some authors believe that CIDN is a rare form of congenital melanocytic nevus due to the clinical and histological similarities. Congenital melanocytic nevi are pigmented lesions which arise as malformations of the neuroectoderm and neural elements, following deregulated growth, and arrest of melanocytes during migration from the neural crest to the skin.[10] However, others suggest that

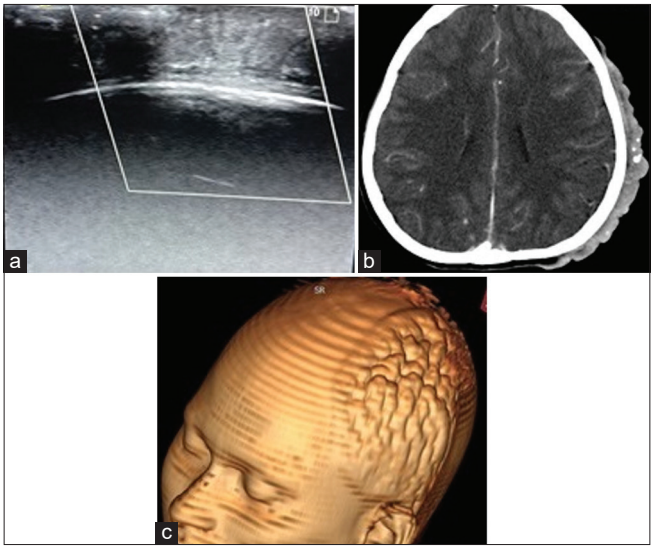


**Figure 3:** (a) Ultrasound examination of the scalp showing thickened skin and subcutaneous tissue with absence of color uptake thus ruling out underlying vascular malformation in a patient with cerebriform intradermal nevus. (b) Contrast-enhanced computed tomography of brain of the same patient showing non-enhancing circumscribed wavy scalp thickening with tiny specks of calcifications involving left parietal region with normal calvarium and brain parenchyma. (c) Contrast-enhanced computed tomography of brain with 3D surface rendering of the same patient showing non-enhancing circumscribed wavy scalp thickening with tiny specks of calcifications involving left parietal region with normal calvarium and brain parenchyma.

CIDN is a separate disorder with a few nevus cell nests at the dermoepidermal junction, associated with alopecia or sparse hair within the lesion.[7,8]

There are also reports of giant congenital nevi with intense pigmentation, increased number of hair follicles, and frequent nevus cell nests at the dermoepidermal junction presenting as CVG.[7,9] As malignant melanoma has been reported in patients with congenital nevus, regular follow-up is mandatory in all cases of CIDN.[10]

Since our patient was very much concerned about the cosmetic appearance of the lesion, a plastic surgical correction was done. She was lost for follow-up after surgery.

## CONCLUSION

Cerebriform intradermal nevus presenting as CVG is a rarity. Histopathology helps to confirm the diagnosis. Ultrasound examination of the scalp and CT brain often help to rule out any underlying vascular malformation.

### Declaration of patient consent

Not required as patients identity is not disclosed or compromised.

### Financial support and sponsorship

Nil.

### Conflicts of interest

Dr. Najeeba Riyaz is on the editorial board of the Journal.

## REFERENCES

1. Yang JJ, Sano DT, Martins SR, Tebcherani AJ, Sanchez AP. Primary essential cutis verticis gyrata-case report. An Bras Dermatol 2014;89:326-8.
2. Lamelas AM, Taub PJ, Silver L, Kressel A. Congenital primary essential cutis verticis gyrata. Eplasty 2016;16:ic13.
3. Diven DG, Tanus T, Raimer SS. Cutis verticis gyrata. Int J Dermatol 1991;30:710-2.
4. Ross JB, Tompkins MG. Cutis verticis gyrata as a marker of internal malignancy. Arch Dermatol 1989;125:434-5.
5. Schöttler L, Denisjuk N, Körber A, Grabbe S, Dissemond J. Asymptomatic cerebriform folds of the scalp. Diagnosis: Primary cutis verticis gyrata (CVG). Hautarzt 2007;58:1058-61.
6. Larsen F, Birchall N. Cutis verticis gyrata: Three cases with different aetiologies that demonstrate the classification system. Australas J Dermatol 2007;48:91-4.
7. Ghosh SK, Bandyopadhyay D, Ghoshal L, Sarkar S. Lobulated mass on the back of the scalp. Indian J Dermatol Venereol Leprol 2012;78:225-7.
8. González JA, Díez MT, Gijón RC, Diaz RM, Olasolo P. Letter: Cerebriform intradermal nevus presenting as secondary cutis verticis gyrata. Dermatol Online J 2010;16:14.
9. van Geest AJ, Berretty PJ, Klinkhamer PJ, Neumann HA. Cerebriform intradermal naevus (a rare form of secondary cutis verticis gyrata). J Eur Acad Dermatol Venereol 2002;16:529-31.
10. Lyon VB. Congenital melanocytic nevi. Pediatr Clin North Am 2010;57:1155-76.

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