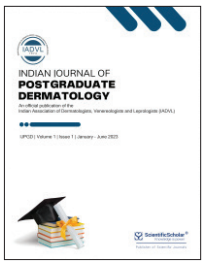
ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

https://ijpgderma.org

Indian Journal of Postgraduate Dermatology



Case Report

Cutis Verticis Gyrata

Sahana Srihari[1], Srihari Iyer[2], Richa Tayal[1], Vippan Goyal[1]

[1]Department of Dermatology, Adesh Institute of Medical Sciences and Research, [2]Department of Aersospace Medicine, Indian Army, Bathinda, Punjab, India.

***Corresponding author:**
Sahana Srihari,
Department of Dermatology,
Adesh Institute of Medical Sciences and Research,
Bathinda, Punjab, India.

drsahanasrihari@gmail.com

Received: 13 February 2024
Accepted: 27 May 2024
EPub Ahead of Print: 01 August 2024
Published: 23 August 2024

DOI
10.25259/IJPGD_24_2024

Quick Response Code:

ABSTRACT

Cutis verticis gyrata (CVG) is a benign dermatological condition characterised by excessive scalp thickening, resembling the gyri of the cerebral cortex and resistant to external pressure. Diagnosis of CVG relies primarily on clinical examination. Additional investigations are warranted to distinguish between primary and secondary forms of CVG and to rule out potential underlying local or systemic aetiologies. This article presents a case report of primary essential CVG alongside unilateral congenital conjunctival melanosis in a 23-year-old male patient.

Keywords: Cutis verticis gyrata, Bulldog scalp, Paquidermia verticis gyrata, Cerebriform intradermal nevus

INTRODUCTION

Cutis verticis gyrata (CVG) represents a benign dermatological anomaly characterised by aberrant scalp growth, resulting in pronounced furrows and intricate folds resembling cortical convolutions of the brain which are refractory to external compression. Diagnosis primarily relies on clinical assessment, with additional investigations often necessary to differentiate primary from secondary CVG and to exclude potential underlying local or systemic pathology. Herein, we present a case of primary essential CVG with unilateral congenital conjunctival melanosis.

CASE REPORT

A 23-year-old male, previously unremarkable in medical history, presented with a chief complaint of a gradually progressing, mildly painful thickening of the scalp characterised by cerebriform convoluted folds and furrows, notably prominent in the parietal and occipital regions, over 6 months, accompanied by mild pruritus. The affected area measured approximately 10 cm in length and 6 cm in width [Figure 1a]. Physical examination revealed absence of signs indicative of infectious or inflammatory processes. Texture of scalp was soft on palpation, with non-tender folds and ridges resistant to traction [Figure 1b]. The patient reported difficulties in hairdressing due to sparse hair growth overlying the folds. The patient acknowledged use of unlabelled protein powder as suggested by a fitness instructor and denied taking any anabolic steroids. There was no history of neurological abnormalities, seizures or head trauma, although the patient experienced occasional headaches. In addition, since birth, the patient has exhibited unilateral conjunctival melanosis without associated visual impairment, although recent complaints of ocular discomfort were noted [Figure 2]. Family history was unremarkable.

A scalp punch biopsy revealed normal scalp architecture with notable increases in collagen density and dermal matrix size, alongside sebaceous hyperplasia, without evidence of

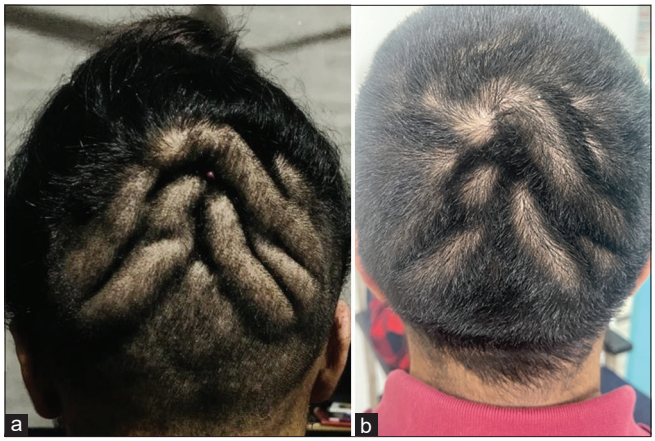


Figure 1: (a) Area of abnormality was 10 cm long and 6 cm wide. (b) The scalp was soft on palpation, folds were non-tender and ridges did not resolve with traction.

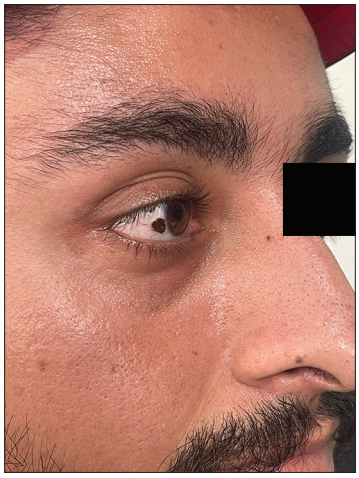


Figure 2: Conjunctival melanosis.

inflammatory or neoplastic pathology. Comprehensive haematological and biochemical investigations, including assessment of serum testosterone, insulin, blood sugar, thyroid function and growth hormone levels, yielded results within normal limits. Negative findings were also obtained for antinuclear antibodies and syphilis screening tests. Systemic examination revealed no abnormalities. Magnetic resonance imaging studies demonstrated no cranial or cerebral anomalies beyond the thickening of the scalp and subcutaneous tissue. Based on these findings, the diagnosis was consistent with primary CVG. The patient was counselled about the condition and educated on proper scalp hygiene practices. Surgical intervention was discussed but deferred at present.

DISCUSSION

CVG, first described in 1837 by Alibert, is also referred to as paquidermia verticis gyrata or cutis verticis plicata. It is said to affect approximately 1 in 100,000 men and 1 in 400,000 women.[1]

CVG is categorised based on its aetiology, with cases lacking identifiable causes termed primary essential CVG. Patients with this form typically present without underlying ocular or neurological pathology. Conversely, primary non-essential CVG, the most prevalent variant, is associated with neurological and ocular conditions, and occasionally, auditory impairments.[2] It often co-occurs with intellectual disability, neuropsychiatric disorders, seizures and ophthalmologic abnormalities. These patients commonly exhibit low IQ, microcephaly and convulsions, although these conditions do not directly precipitate scalp folding.[1] Chromosomal aberrations such as fragile X syndrome, Turner syndrome and Klinefelter syndrome may be implicated.[3]

In primary CVG, furrows typically align in an antero-posterior orientation, with creases and raised areas evenly distributed, often observed at the apex and occiput. This variant predominantly affects males before the age of 30, leading to speculation regarding a hormonal aetiology, possibly related to serum testosterone levels. The precise relationship between testosterone and CVG remains enigmatic, although associations with collagen thickening and sebaceous gland hypertrophy have been noted.[4]

Secondary CVG arises from underlying defects directly causing scalp folds and furrows. This form can result from neoplastic processes, systemic disorders or medication use, such as anabolic steroids.[5] Reported associations include pachydermoperiostosis (idiopathic hypertrophic osteoarthropathy), cerebriform intradermal nevus and acromegaly, among others. Secondary CVG can manifest at any age, without gender predominance, with fold direction correlating with the underlying disorder.

Histopathological examination of primary CVG may reveal a spectrum of findings, ranging from normal skin architecture to thickened connective tissue with hypertrophy and hyperplasia of adnexal structures, potentially accompanied by increased collagen fibres and entrapment of apocrine and eccrine glands. Conversely, secondary CVG typically exhibits histopathological features indicative of the underlying disease process. It is imperative to exclude local or systemic causes that may predispose to secondary CVG through comprehensive complementary investigations before conclusively diagnosing primary CVG.[6]

Maintaining meticulous local scalp hygiene is essential to prevent the accumulation of secretions, maceration, malodour and subsequent secondary infections. Addressing the underlying condition in cases of secondary CVG typically leads to regression of the CVG presentation. Management

of primary CVG often centres on addressing psychological or cosmetic concerns. Surgical intervention, ranging from straightforward excision to more complex procedures such as skin grafting, remains the preferred treatment modality. The prognosis for CVG is generally favourable, as there is no documented evidence of malignant transformation in the literature.

CONCLUSION

CVG is a benign disorder. Yet, an interdepartmental approach by dermatologists, paediatricians, surgeons, internists, etc., is essential to get the best outcomes. The management of primary CVG and its secondary associations in a holistic manner will improve the outcomes and decrease morbidity and lead to optimal results. Family education regarding the disorder is also much needed.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Shareef S, Horowitz D, Kaliyadan F. Cutis Verticis Gyrata. In: StatPearls. Treasure Island, FL: StatPearls Publishing;2024.
2. Sriphrapradang C, Ngarmukos C. Bulldog Scalp. Cleve Clin J Med 2016;83:90-1.
3. Larsen F, Birchall N. Cutis Verticis Gyrata: Three Cases with Different Aetiologies that Demonstrate the Classification System. Australas J Dermatol 2007;48:91-4.
4. Cohen B, McKenzie Maloney BS. Scalp Thickening and Folding in a Pubertal Boy. 2023.
5. Kim HS, Teo RY, Tan AW. Cutis Verticis Gyrata in a Patient with Hyper-IgE Syndrome. Acta Derm Venereol 2009;89:413-4.
6. Sankarapandian J, Palaniappan V, Karthikeyan K. Primary Essential Cutis Verticis Gyrata: A Case Report with a Review of Literature. Indian Dermatol Online J 2023;15:313-5.

How to cite this article: Srihari S, Iyer S, Tayal R, Goyal V. Cutis Verticis Gyrata. Indian J Postgrad Dermatol. 2024;2:126-8. doi: 10.25259/IJPGD_24_2024