A Curious Case of 'Brain-Like Scalp'

[1]**Dr. Sneha Arakeri**

Post-graduate, MD Dermatology Venerology and Leprosy

KLE University, JNMC Medical College

[2]**Dr. Shivakumar Patil**

Asso. Professor, MD Dermatology Venerology and Leprosy

KLE University, JNMC Medical College

Introduction

Cutis verticis gyrata (CVG) is a rare cutaneous disorder first reported in 1837 by Alibert.[1] It is also known as bulldog scalp syndrome, cutis verticis plicata, corrugated scalp, and pachydermia verticis gyrata. The condition is classified into primary and secondary forms. The primary form has two subtypes: essential (without associated comorbidities) and non-essential (with neurological or ophthalmological associations).[2] The secondary form results from underlying pathology such as inflammatory conditions, neoplasms, or connective tissue disorders.[3] CVG predominantly affects males and is often diagnosed in post-pubertal individuals.[1]

Case Report

A 29-year-old male presented to the dermatology outpatient department with a history of uneven thickening of the scalp, first noticed one month prior. The patient reported no changes in size or shape of the thickening, and no associated symptoms such as fever, visual disturbances, seizures, speech difficulties, behavioral changes, or intellectual impairment. He had no history of comorbidities, was not on long-term medication, and had no family history of similar conditions. On examination, multiple folds with deep furrows were observed over the occipital and vertex regions of the scalp (figure 1). The overlying skin appeared normal, without signs of inflammation, tenderness, or temperature changes. Histopathological examination revealed localized hyperkeratosis, thickened collagen fibers, areas of hemorrhage, and a chronic inflammatory infiltrate in the upper dermis (figure 2).

Discussion

CVG is classified into primary and secondary types. Primary CVG occurs in the absence of an identifiable underlying cause and can be further divided into essential and non-essential types. The essential type has no associated systemic conditions, whereas the non-essential type may be linked to neurological and ophthalmological abnormalities, such as epilepsy, cerebral palsy, mental retardation, cataracts, and retinitis pigmentosa [1]. The secondary form, on the other hand, arises due to underlying systemic conditions, including acromegaly, pachydermoperiostosis, and inflammatory diseases of the scalp [2].

Several theories have been proposed regarding the pathogenesis of CVG. One hypothesis suggests that hypertrophy of the scalp tissues, possibly due to excessive production of collagen and fibroblast proliferation,

leads to the characteristic thickened folds [3]. Another theory implicates hormonal influence, as CVG is more commonly observed in post-pubertal males, hinting at a possible role of androgens in its development [1]. The folds in primary CVG are usually symmetrical and non-tender, whereas secondary CVG may present with additional symptoms depending on the underlying disease.

Histopathological analysis plays a crucial role in distinguishing CVG from other dermatological and systemic conditions. Findings typically include epidermal hyperkeratosis, thickened dermal collagen, and perivascular inflammatory infiltrate. Biopsy and histopathological examination are essential in ruling out secondary causes [2].

The diagnosis of primary essential CVG is largely clinical, with histopathology serving as a confirmatory tool. Differential diagnoses include acromegaly, cutis laxa, cerebriform intradermal nevus, and inflammatory scalp conditions. Management is primarily conservative, focusing on aesthetic concerns, but surgical correction may be considered in severe cases. Some cases have been treated with scalp reduction surgery or tissue expansion techniques to remove the excess folds and improve cosmetic appearance [3].

Conclusion

This case highlights the importance of recognizing CVG and distinguishing between its primary and secondary forms. While the condition is benign, its impact on aesthetics and potential associations with systemic diseases warrant a thorough evaluation. Future studies should explore optimal treatment strategies, including surgical and non-surgical approaches, to improve patient outcomes.

References

1. Shareef S, Horowitz D, Kaliyadan F. Cutis Verticis Gyrata. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan–. PMID: 30969634.

2. Ennouhi M, Guerrouani A, Moussaoui A. Idiopathic Cutis Verticis Gyrata in a Female. Cureus. 2018 Jan 23;10(1):e2105. doi:10.7759/cureus.2105.

3. Yang JJH, Sano DT, Martins SR, Tebcherani AJ, Sanchez APG. Primary essential cutis verticis gyrata - Case report. An Bras Dermatol. 2014;89(2):326-8.

Legends

Figure 1: Clinical image of scalp showing multiple folds with deep furrows over the occipital and vertex regions

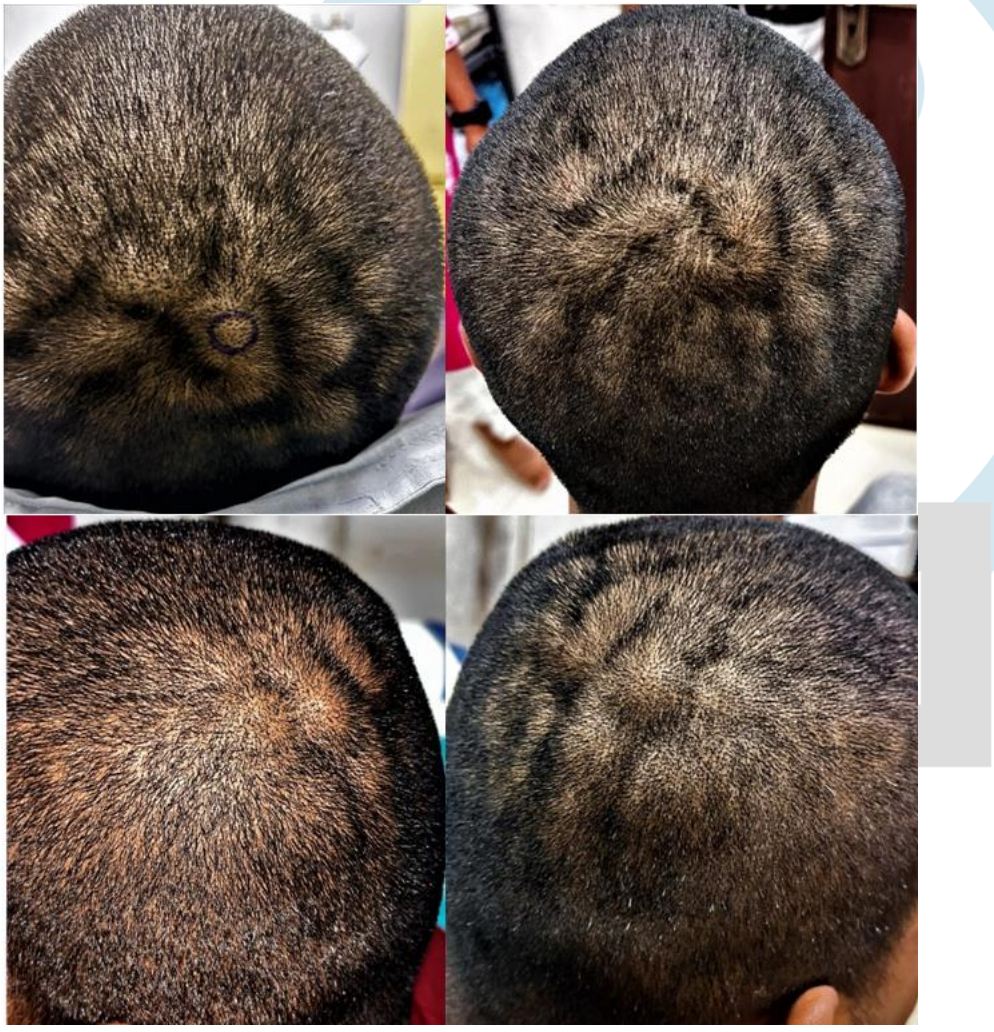

Figure 2: 40x HE stained section showing localized hyperkeratosis, thickened collagen fibers, areas of hemorrhage, and a chronic inflammatory infiltrate in the upper dermis