Primary Essential Cutis Verticis Gyrata: A Case Report

Cutis Verticis Gyrata Essencial Primário: Relato de Caso

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Palavras-chave: Anomalias da Pele; Couro Cabeludo; Dermatoses do Couro Cabeludo

Dear Editor,

Cutis verticis gyrata (CVG) is a rare scalp condition characterized by excessive subcutaneous tissue proliferation, resulting in folds resembling cerebral gyri.[1] First documented by Jean-Louis-Marc Alibert in 1837 and later named by Paul Gerson Unna in 1907, it is also known by various names like Robert-Unna syndrome, bulldog scalp, and corrugated skin. The estimated prevalence rate in adults is 1 in 100 000 for men and 0.026 in 100 000 for women.[2]

CVG is categorized into primary and secondary forms. Primary CVG can be further divided into essential (unrelated to any other disease) and non-essential (associated with neurological abnormalities like microcephaly, mental deficiency, and ophthalmological conditions). Secondary CVG is linked to underlying systemic disorders, endocrine conditions, inflammatory dermatoses, neoplasms, or trauma.[3]

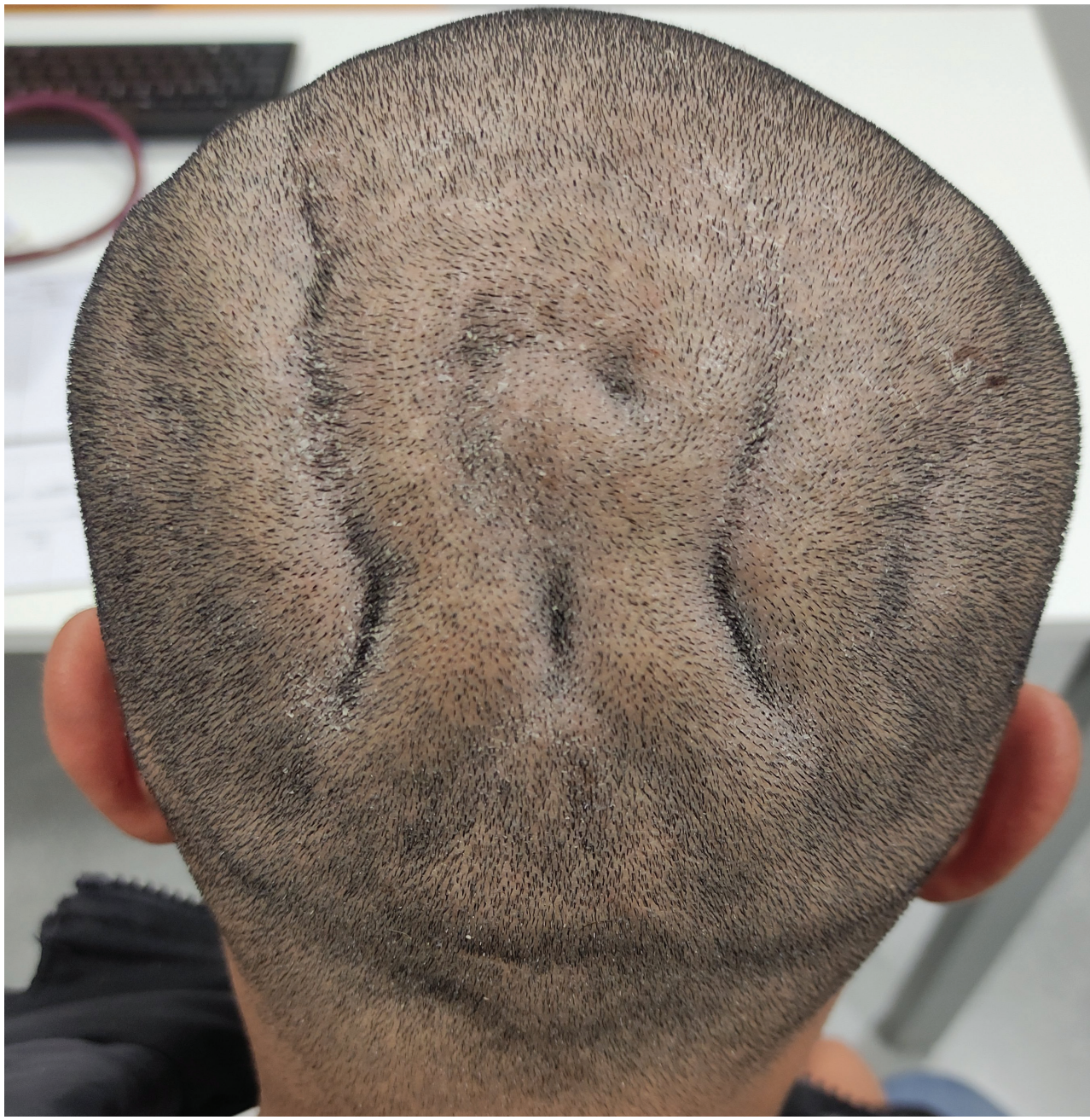

Figure 1 – Posterior picture of the scalp showing 4 - 5 prominent furrow-lines without concomitant lesions

In this case report, a 22-year-old man without prior medical history admitted in the emergency room presented scalp lesions that had developed over two months without other symptomatology (Fig. 1). The physical examination revealed prominent furrow-like folds on the scalp with no other skin lesions or associated symptoms. All secondary causes were excluded, including endocrinological diseases such as acromegaly due to the normal values of IGF-1 and GH. The biopsy showed slight acanthosis, elongation of the epidermal ridges and slight hyperkeratosis in the epidermis. In the superficial dermis, the biopsy showed a perivascular lymphohistiocytic infiltrate, vascular ectasia and slight perifolliculitis.

The patient was diagnosed with primary essential CVG and remained under surveillance at the Dermatology clinic. Surgical intervention was considered if the lesions increased in size causing psychological distress or if there were recurrent infections.[2]

The clinical presentation of CVG resembles cerebral folds and furrows in the scalp. Primary essential CVG has no known cause, but it is theorized that autosomal dominant mutations in fibroblast growth factor receptor 2 could be related. Other forms of CVG may be linked to various underlying conditions, including hormonal imbalances, inflammatory skin diseases, infectious diseases, and tumors.[4]

Diagnosing CVG is primarily based on clinical appearance, characterized by 2 to 30 folds measuring 0.5 to 2 cm in width and around 1 cm deep, typically found in the occipital and vertex regions of the scalp.[1] Skin biopsies can help differentiate primary CVG from secondary forms. However, this diagnostic tool may not help in some situations, including acromegaly, where no specific histological changes are observed. The histopathological findings on primary CVG will reveal hypertrophy and hyperplasia of epidermal appendages and thickened dermal collagen in the skin, whereas the histopathological changes in secondary CVG include abnormalities that are characteristic of the underlying etiology.[5] Treatment depends on the severity and patient preference, ranging from surveillance for stable or mild cases to surgical options like excision, grafting, or tissue expansion for those with psychological distress, cosmetic concerns or recurrent infections.[2]

AUTHOR CONTRIBUTIONS

All authors contributed equally to this manuscript.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in 2013.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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REFERENCES

1. Zeng W, Guo L. A rare cutis verticis gyrata secondary to cerebriform intradermal nevus: case report and literature review. BMC Surg. 2021;21:234.
2. Snyder MC, Johnson PJ, Hollins RR. Congenital primary cutis verticis gyrata. Plast Reconstr Surg. 2002;110:818-21.
3. Lamelas AM, Taub PJ, Silver L, Kressel A. Congenital primary essential cutis verticis gyrata. Eplasty. 2016;16:ic13.
4. Tucci A, Pezzani L, Scuvera G, Ronzoni L, Scola E, Esposito S, et al. Is cutis verticis gyrata-intellectual disability syndrome an underdiagnosed condition? A case report and review of 62 cases. Am J Med Genet A. 2017;173:638-46.
5. Chamli A, Jones M, Bacha T, Litaiem N, Zeglaoui F. Cutis verticis gyrata: three cases illustrating three different etiologies. Clin Case Rep. 2022;10:e05889.

Eduardo ESTEBAN-ZUBERO✉[1], Cristina GARCÍA-MURO[2]
1. Emergency Department. Hospital San Pedro. Logroño. Spain.
2. Department of Pediatrics. Hospital San Pedro. Logroño. Spain.
✉ **Autor correspondente:** Eduardo Esteban-Zubero. eezubero@gmail.com
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