

Primary essential cutis verticis gyrata: A case report

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ABSTRACT

Cutis verticis gyrata (CVG) is a rare condition of the scalp characterized by convoluted folds and furrows produced by the excessive growth of the skin of the scalp and resembling the cerebral gyri. CVG can be identified as primary—essential or nonessential—or secondary. Herein, we report the case of a 20-year-old female with primary essential CVG, who presented herself with thickened and convoluted skin folds over the vertex and parietal region of the scalp persistent for one year prior without other symptoms. CVG is a rare congenital or acquired disease distinguished by redundancy of the scalp skin that resembles the cerebral gyri. The management of primary essential CVG can be symptomatic or surgical depending on the patient's aesthetic expectations.

Key words: Cutis verticis gyrata; Scalp dermatoses; Rare

INTRODUCTION

Cutis verticis gyrata (CVG) is a rare disease characterized by the excessive growth of the skin of the scalp, resulting in furrows and folds that resemble the gyri of the brain cortex [1,2]. Depending on its form, CVG can be identified as primary—essential or nonessential—or secondary [2]. Primary nonessential CVG is associated with neurological and ophthalmological manifestations while primary essential CVG is not. The secondary form of CVG is much more frequent and accompanies other pathologies [3]. There have been around 28 cases of primary essential CVG described in the literature. We report a case of primary essential CVG in a young female.

CASE REPORT

A 20-year-old female presented herself with a history of thickened and prominent skin folds over the vertex and parietal region of the scalp persistent for one year prior (Fig. 1). No other symptoms were reported. A physical examination revealed enlargement of the skin folds of the scalp arranged in a vertical position.

There was no history of comorbidity, no family history of similar lesions, and no history of consanguinity. The

patient reported no neurological or ophthalmological symptoms. Neurological, psychiatric, ophthalmological, and endocrine consultations were obtained and, to exclude secondary causes, a complete blood count, a thyroid function test, and syphilis screening were done and the levels of growth hormone, follicle-stimulating hormone, luteinizing hormone, and serum cortisol were checked, all of which reported normal values. MRI of the brain revealed a thickened scalp with marked ridges and furrows of a gyriform appearance in the parietooccipital region, suggestive of cutis verticis gyrata with no underlying defects. As the clinical picture was pathognomonic of CVG, a skin biopsy and histopathology were deemed unnecessary.

DISCUSSION

Cutis verticis gyrata (CVG), also known as cutis sulcata, cutis capitis strata, cutis verticis plicata, or the “bulldog scalp,” was first described in 1837 by Jean-Louis-Marc Alibert, and the term *cutis verticis gyrata* was proposed by Unna in 1907 [1,4]. Depending on its form, CVG can be identified as primary or secondary (Fig. 2) [3].

Primary CVG refers to cases with no underlying cause [4], beginning after puberty, usually before

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the age of thirty, and occurring predominantly in males, with a male-to-female ratio of 5:1. Primary CVG is divided further into two types: essential and nonessential (Fig. 2) [4,5].

The essential form, exhibited by our patient, is a rare disorder characterized by the thickening of the scalp, where the excess skin folds are symmetrical and take the shape of the cerebral gyri. There is no underlying neurological or ophthalmological association [1].

The nonessential form is associated with neurological (epilepsy, intellectual disability, microcephaly), psychiatric (schizophrenia), and ophthalmic (cataract, strabismus, blindness, retinitis pigmentosa) disorders [1,4].

Secondary CVG is more common than primary CVG, can occur at any age, and displays no gender predilection. Secondary CVG has a number of underlying causes [5]. Several sources explain that the secondary form may

develop after inflammatory dermatosis, endocrine and genetic disorders, and internal malignancies as paraneoplastic syndrome (Table 1) [1,6].

CVG usually affects the vertex and the occipital scalp, and may sometimes involve the entire scalp. Generally, the folds are arranged in an anteroposterior direction but may be transverse on the occiput [5]. CVG is usually asymptomatic but the patient may sometimes complain of pruritus, a burning sensation, malodor, and thinning of the hair in the folded areas due to aggregation of debris and skin secretions in the folds [7].

The diagnosis of CVG is based on clinical findings but investigations are necessary to distinguish between primary and secondary CVG [5].

The treatment of primary essential CVG depends on the patient's aesthetic expectations, since no organic function



Figure 1: Scalp hypertrophy with anteroposterior folds in the vertex and occipital region of the head.

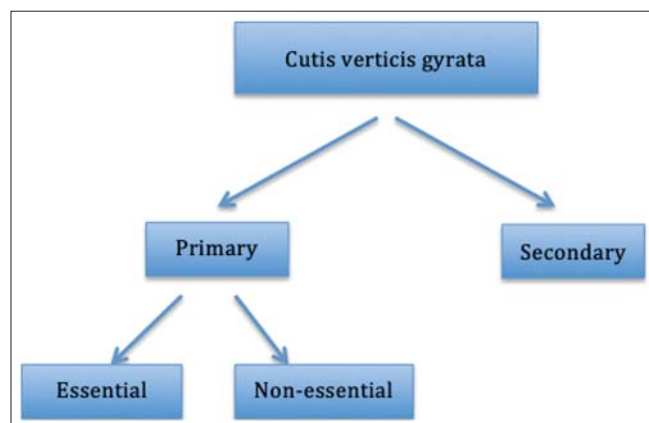


Figure 2: The classification of cutis verticis gyrata (CVG).

Table 1: Diseases associated with CVG

Type of CVG	Associated conditions
Primary [1,6]	
Essential	No associated abnormalities
Nonessential	Mental deficiency, cerebral palsy, seizures, microcephaly, ophthalmological abnormalities (cataract, optic atrophy, strabismus, blindness, retinitis pigmentosa)
Secondary [1,4,6,7]	(local underlying process, usually asymmetric, onset at any age)
Infectious diseases	Folliculitis, erysipelas, impetigo, syphilis
Inflammatory disorders	Eczema, scalp psoriasis, keloidal acne
Inherited diseases	Klinefelter syndrome Turner syndrome Noonan syndrome Ehlers–Danlos syndrome Michelin tire baby syndrome Fragile X syndrome Beare–Stevenson syndrome Tuberous sclerosis
Tumors	Neurofibromatosis Cylindroma Lymphangioma Leukemia Pituitary tumor Intracerebral aneurysm Dermatofibroma
Nevi	Melanocytic nevus Lipomatous nevus Sebaceous nevus Cerebral intradermal nevus
Endocrine	Acromegaly Myxedema Diabetes mellitus type 2
Miscellaneous	Drugs (anabolic steroids) Chronic traction Acanthosis nigricans Trauma Acne conglobata Pachydermoperiostosis Amyloidosis Mucinosis

needs to be corrected. Local hygiene is important to prevent the accumulation of secretions with an unpleasant odor [8]. Surgical treatment may be considered to remove excess skin folds with the aim to achieve a more aesthetically satisfying appearance of the scalp. In our case, CVG was easily concealed by the patient's long hair. She was counseled regarding the benign nature of her condition and informed about the possibility of secondary infection due to the trapping of debris in the depressions. Explained to her was also the importance of local hygiene and attending regular follow-ups.

CONCLUSION

Primary essential CVG is an extremely rare condition, with around 253 cases of CVG and 28 cases of primary essential CVG reported in the literature. Because it is a rare condition, our case appears as a valuable educational opportunity. A dermatologist finding cases such as ours should take a proper medical history and perform careful examinations and complementary investigations to reach a definitive diagnosis.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The

patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

REFERENCES

1. Yang JJH, Sano DT, Martins SR, Tebcherani AJ, Sanchez APG. Primary essential cutis verticis gyrata - case report. *An Bras Dermatol*. 2014;89:326-8.
2. Zeljko-Penavić J, Šimić D, Jurišić K, Gunarić A. Cutis verticis gyrata - a case report. *Acta Dermatovenerol Croat*. 2016;24:140-3.
3. Walia R, Bhansali A. Cutis verticis gyrata. *BMJ Case Rep*. 2011;10.1136/bcr.01.2011.3763.
4. Er O, Külcü Çakmak S, Kayaçetin S, Tamer E, Artüz F. Cutis verticis gyrata secondary to cerebriform intradermal nevus. *Our Dermatol Online*. 2016;7:406-8.
5. Farrant P, Mowbray M, Sinclair RD. Dermatoses of the scalp. In: Griffiths C, Barker J, Bleiker T, Chalmers R, Creamer D, editors. *Rook's textbook of dermatology*. 9th ed. London: John Wiley and Sons, Ltd; 2016; Vol 3 p. 107.9.
6. Mohammadi S, Khalili M, Mohebbi A, Afatoonian M, Badakhsh H. Cutis verticis gyrata and leonine face in a patient with Darier disease: a case report and review of literature. *J Pak Assoc Dermatol*. 2018;28:114-6.
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. Elharrouni AA, Douhi Z, Elloudi S, Baybay H, Mernissi FZ. Primary essential cutis verticis gyrate. *Our Dermatol Online*. 2020;11(e):e14.1.

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