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Cutis verticis gyrata: Clinical image

Ana Paula Santos de Assis, Izabela Schneidewind, Maria Laura Fernandes Cordeiro, Antônio Marcos da Silva Catharino

CASE REPORT

Male patient, 37 years old, was brought to the neurology clinic due to the appearance of the scalp.

The patient had a history of chronic non-progressive encephalopathy, resulting from congenital rubella. He was diagnosed with microcephaly at birth and, later, craniostenosis and epileptic seizures. He underwent corrective surgery for congenital malformation. Today, mental retardation and tetraparesis are observed. He currently uses phenobarbital 100 mg/day, getting reduction of the seizures.

After clinical evaluation, skin folds, which resembled the appearance of the cerebral cortex, were observed on the scalp. The patient was diagnosed with primary non-essential cutis verticis gyrata (CVG) (Figure 1).

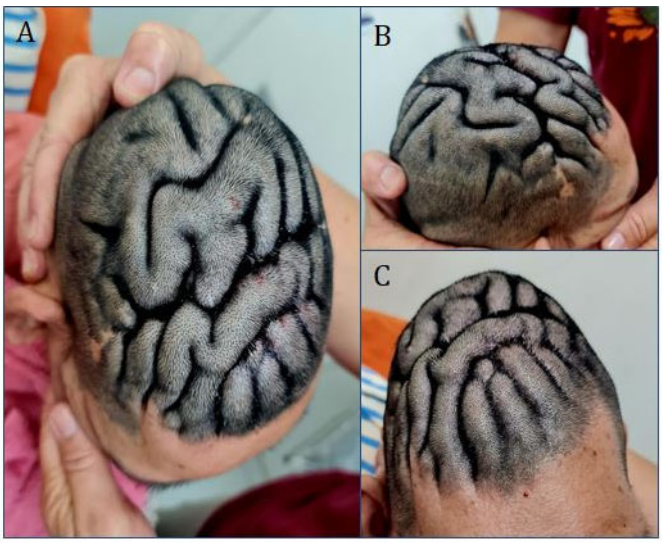

Figure 1: Skin folds in the scalp. (A) View from above; (B) Right side view; (C) Anterior view.

DISCUSSION

The CVG is a rare and benign dermatological disease, with a prevalence of 0.026–1 affected in 100,000 live births, commonly found in males after puberty. Roughly speaking, it may be known as “Bulldog Scalp Syndrome” [1].

It is characterized by excessive and abnormal hyperplasia and hypertrophy of the skin of the scalp, causing deformities and redundant formation of grooves that resemble the gyres of the cerebral cortex [2]. As a result of this change, there is an increase in tissue thickness, circumvolutions, and multidirectional folds in cerebriform shape.

The first report of this disease in bibliography is carried out by Jean-Louis-Marc Alibert, in 1837, who called it cutis sulcata [3]. As early as 1907, the disease was given a second nomenclature by Paul Gerson Unna who gave it other names such as Robert-Unna syndrome, bulldog scalp, corrugated skin, cutis verticis plicata, and pachydermia verticis gyrata.

These skin changes characterized by folds are visible and can vary in quantity, thickness, and consistency, and are typically spongy and often affect the occipital region. Although it is a subclassified disease, all its presentations involve a change in the distribution of the scalp [4].

The pathophysiological process characteristic of the disease is still unclear, but theories suggest the emergence due to excessive deposition of collagen on the dermis of the scalp, which causes thickening. Others also estimate that, due to the higher incidence of the disease at puberty, there is hormonal influence in its occurrence. Regardless of the theory adopted, it is possible that cutis verticis gyrata is an autosomal inheritance [5].

Ana Paula Santos de Assis[1], Izabela Schneidewind[1], Maria Laura Fernandes Cordeiro[2], Antônio Marcos da Silva Catharino[3]

Affiliations: [1]Medical student at Universidade Iguaçu, Campus I and effective member of the Academic League of Neurology, UNIG/Nova Iguaçu, RJ, Brazil; [2]Medical student at Universidade Iguaçu Campus I, UNIG/Nova Iguaçu, RJ, Brazil; [3]Department of Neurology of Hospital Geral de Nova Iguaçu, PhD student in Neurology at the Federal University of the State of Rio de Janeiro - UNIRIO, Adjunct Professor of Medicine at Iguaçu University - UNIG/Nova Iguaçu, RJ, Brazil.

Corresponding Author: Antônio Marcos da Silva Catharino, Rua Gavião Peixoto 70, Room 811, CEP 24.2230-100, Icaraí, Niterói-RJ, Brazil; Email: catharino.antonio@gmail.com

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The histopathology shows sebaceous hyperplasia, and there may be an increase in collagen, not occurring malignant transformation of the skin, nor the cerebral parenchyma.

Similar to the pathophysiology, the etiology is still unknown, but it is known that it is not an exclusively congenital condition, and can be classified as primary, essential, or non-essential, and secondary, being related to a diversity of diseases, inflammatory, endocrinological, paraneoplastic, hematological, or iatrogenic pathological processes [6].

The primary non-essential form is responsible for 0.5% of patients with cognitive impairment. In addition, microcephaly, schizophrenia, cerebral palsy, deafness, epilepsy, cataract, and amaurosis may be present [7]. Contrary to this manifestation, the primary essential form is not associated with neurological and ophthalmologic changes occurring only the formation of folds in the scalp. Here, the folds are usually found symmetrically.

Secondary forms of CVG are represented by pachydeoperiostosis, acromegaly, amyloidosis, syphilis, underlying malignancy and intradermal cerebriform nerve [8]. Secondary forms of the disease occur frequently in both sexes, presenting in different age groups and may result from inflammatory processes or other previous diseases [9, 10]. Here, the folds are found asymmetrically.

Other associated medical conditions include inflammatory diseases of the scalp (eczema, psoriasis, folliculitis, impetigo, erysipelas, and Pênfigos), myxedema, leukemia, syphilis, Acanthosis nigricans, tuberous sclerosis, Ehlers–Danlos syndrome, amyloidosis, diabetes mellitus, pruritus, bleeding, secondary infections, and foul odor due to bacterial proliferation in the circumvolutions.

For diagnostic confirmation, it is necessary to rule out secondary changes and, from the clinical evaluation and completion of complementary tests, such as magnetic resonance imaging, electroencephalogram, cranial computed tomography, microscopy, neurocognitive tests, and laboratory tests (blood count, hepatic enzymes, alkaline phosphatase, creatinine, urea, calcium, free T4 and TSH), discard other probable pathologies [11].

The management of the disease may include conservative management with correct asepsis of the folds areas, as well as surgery, if requested for psychological or aesthetic reasons, and treatments available for basic diseases [12]. In addition, psychological counseling and outpatient follow-up for symptomatic management are essential.

The surgical treatment is performed in two times, where in the first surgery tissue expanders are used in the cranial cap previously, for the preparation of the skin, which allows greater tissue gain for the second surgical time, surgical wound resection and closure after excision, which is still controversial [13, 14].

The aesthetic aspect is relevant due to the extensive area it occupies. Therefore, different techniques are used to reduce scalp defects, such as total resection of the lesion and grafting; use of skin expander in the healthy area and grafting; partial resection of the most abundant part of the lesion.

Usually, the affected area is asymptomatic; however, there may be accumulation of secretions between the folds, causing unpleasant odor and itching sensation, eventual skin infections, mainly fungal, due to the difficult hygiene of the furrows.

There should be counseling in relation to the disease for the affected patient, from a multidisciplinary therapy, with collaboration between plastic surgeons, pathologists, dermatologists, psychiatrists, and psychologists; total resection of the lesion implies definitive alopecia, with possible emotional damage to the patient.

CONCLUSION

Based on the progression of the disease, the importance of early diagnosis is highlighted, informing patients about the damage caused by it. In this case report, it is possible to evaluate that the patient presents the diagnosis of primary non-essential cutis verticis gyrata due to the presence of neurological changes. In addition, through its symptomatology, although the effective response to phenobarbital, it is worth emphasizing the need for constant and multidisciplinary outpatient follow-up, aimed at reducing damage and emotional repercussions to the patient and/or his family.

Keywords: Bulldog scalp syndrome, Cutis verticis gyrata, Scalp skin hypertrophy

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Author Contributions

Ana Paula Santos de Assis – Conception of the work, Design of the work, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Izabela Schneidewind – Conception of the work, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Maria Laura Fernandes Cordeiro – Conception of the work, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Antônio Marcos da Silva Catharino – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.