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Cútis Vértice Gyrata - A Case Report

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Abstract: <u>Introduction</u>: Cutis Verticis Gyrata (CVG) is a rare skin disease, caused by overgrowth of the scalp, cerebriform fashions and wrinkles. CVG can be classified into two forms: primary (essential and non-essential) and secondary. The primary non-essential form is associated with neurological and ophthalmological abnormalities, whereas the primary non-essential form occurs without associated comorbidities. <u>Case report</u>: We report a rare case of primary essential CVG with a history of asymptomatic, normal-colored scalp skin folds located in the parietal-occipital region in a 28-year-old male patient. Major CT clinics include thickened dermis and scalp overgrowth, forming the characteristic folds of the scalp. <u>Conclusion</u>: Not all patients with CVG essential primary surgical intervention, and clinical observation should be adequate for those in stable condition.

Keywords: Scalp dermatoses; cutis verticis gyrata; skin disease

1. Introduction

Cutis verticis gyrata (CVG), also known as pachyderma verticis gyrata, cutis verticis plicata and bulldog scalp syndrome¹, is a rare benign skin disease characterized by convoluted folds and deep creases of the scalp that mimic cerebral sulci and whirl. It was first reported in 1843, but the term gyrata vertex cutis was introduced by Unna in 1907. The gyrata vertex cutis can be primary or secondary². The etiology of CVG is unknown. It can be primary (non-essential and essential forms) or secondary³

Primary non-essential cutis verticis gyrata may be associated with neuropsychiatric and ophthalmological abnormalities. This form was renamed cutis verticis gyrata-intellectual disability (CVG-ID). Secondary and primary non-essential cutis verticis gyrata have associated abnormalities, whereas primary essential CVG has no association. This activity reviews the etiology, presentation, assessment and treatment of cutis verticis gyrata and examines the role of the interprofessional team in the assessment, diagnosis and treatment of the disease ⁴.

The objective was to describe the case of a patient with cutis verticis gyrata and make a brief review on the subject.

2. Case Presentation

A 28-year-old patient (Fig 1) presented the chief complaint of generalized hair loss in the parietaloccipital region of the scalp for a period of 1 year and the hair loss area with mild itching. In addition, he had skin folds on the scalp that resemble the crest and sulcus of the cerebral cortex in the parietaloccipital region, beginning in adolescence. Physical examination revealed hypertrophy and the formation of folds in the parietal-occipital region, forming 4 grooves. The size of the cerebriform mass was about cm × 8.0 cm, with no other skin lesions. Cranial X-ray: normal structure of the bones of the skullcap, sella turcica with normal appearance. - CT skull: no changes. - Pathology the epidermis presentes irregular acanthosis and hyperkeratosis, in addition to some hair follicles with infundibulums dilated. The dermis showed a discreet infiltrate lymphohistiocytic inflammatory, with some associated eosinophils, distribution perivascular and perifollicular has not been observed signs of neoplasia in the examined sample. This is a rare case of essential primary gyrata vertex cutis.

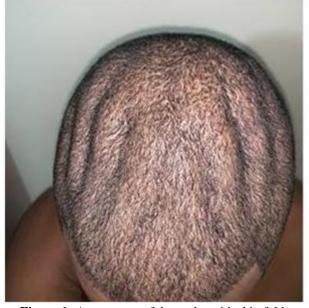


Figure 1: Appearance of the scalp, with skin folds

3. Discussion

Cutis verticis gyrata is a pathology whose clinical diagnosis is usually easy, but the great challenge is to accurately diagnose the underlying or associated conditions⁵. The pathogenesis is unclear, however, a complete classification system is necessary, as CVG is a manifestation from a variety of diverse causes, and is not an individual disease entity. This finding leads to the need to understand the etiology of CVG, for better clinical-surgical management.

Cutis verti cis gyrata is divided into three categories: primary, non-essential and secondary. Primary essential CVG has no other associated abnormalities. Non-essential primary CVG is associated with intellectual disability, neuropsychiatric disorders, seizures, schizophrenia, cerebral palsy, ophthalmological abnormalities such as cataracts, strabismus, retinitis pigmentosa and blindness⁶.

The diagnosis of primary CVG is made after further investigation and as local or systemic causes that cause cutis verticis gyrata secondarily have been excluded Corresponding investigations such as skin biopsies, blood tests, and radiological examinations are recommended based

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on the patient's clinical presentation to distinguish between the primary and secondary forms of CVG⁸. Histopathology shows sebaceous hyperplasia, with no evidence of collagen enlargement 9

The hormonal cause of cutis verticis gyrata has been postulated due to male predominance it usually appears during or after puberty, 90% of patients being diagnosed after age 30 years⁴, and the post-pubescent onset of this disease. However, a study of plasma levels. The levels of thyroid hormone, cortisol, sex hormone and prolactin in 15 patients with cutis verticis girata were not detectable as high levels of abnormal hormones and it was also not possible to prove the association with genetic transmission ⁵.

The primary essential form presents itself only as scalp folds, which mimic brain turns and predominantly affect men;

The treatment starts with good local hygiene of the scalp, which is important to avoid the accumulation of secretion, maceration and to prevent secondary contamination 10

Despite being a benign pathology, a primary cutis verticis gyrata generates changes in the quality of life of patients with psychological and aesthetic repercussions. Continuous surgical intervention is the most effective treatment option.⁵

Secondary cutis verticis gyrata is pachydermoperiostosis (idiopathic hypertrophic osteoarthropathy), cerebriform intradermal acromegaly, and other less frequent causes of CVG, such as inflammatory scalp diseases, scleromyxedema, leukemia, lymphoma, Ehlers' syndrome, diabetes mellitus, cutis laxa, intracerebral aneurysm, focal cutaneous mucinosis and misuse of anabolic steroids¹².

The complications found in this pathology are associated with the patients' quality of life, secondary infections in the deeper folds and in relation to surgical intervention. A surgical complication that can occur in smaller creases is a distortion of the eyebrow, eyelid, and scalp line, depending on the location of the creases. Therefore, the excision pattern must be reproducible, as the disease is progressive ¹³.

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