Cutis verticis Gyrata

*A. O. Ogunbiyi, Lanre Falodun

SUMMARY

Objective: To report and discuss Cutis verticis Gyrate, a rare disorder of the scalp which occurs mainly in men.

Method: We report the case of 30 years old man who presented with folds and furrows on his scalp which started at the age of twelve. The folds were asymptomatic, however patient was worried as it appeared to be progressive.

Observations: The patient had convoluted folds and furrows located anterior posterioly on the scalp. There was no associated hair loss or pigmentary changes. Neurological and apthalmological examinations were found to be within normal limits. A diagnosis of primary non essential cutis verticis gyrate was made.

Conclusion: Cutis verticis gyrate is a rare disorder of the scalp due to thickening of the scalp. It may be primary as was in the case of the patient and was not associated with other systemic disorders. It may also present with other associated disorders. This is the first case of cutis verticis gyrate document in the environment.

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INTRODUCTION

Cutis verticis gyrate is a descriptive diagnosis of folding of the scalp that produces a characteristic gyrate pattern. It occurs in two main forms, primary or secondary. The primary type may be essential or non-essential. The essential form is associated with various disorders such as psychiatric, neurological and opthelmological complications.

The non essential type which our patient presented with is not associated with any underlying disorder, but is quite rare. We present a case with cutis verticis gyrate followed by a brief literature review.

CASE REPORT.

A 30year old man presented in the skin clinic with complaints of an unusual wavy appearance of skin folds on his scalp. He claimed he noticed the development of the way appearance when he was about twelve years old, but was worried because the folds appeared to be more prominent as he grew older. There was no associated scalp pain or hair loss. He was other wise in good health. There was no associated underlying chronic illness. There was no similar problem in any member his family. Examination revealed a healthy Youngman, with folds and furrows on the anterior-posterio aspects of his scalp. The furrows were more prominent on the right side (fig 1). Although he had just had a hair cut, there was no evidence of alopecia or differential density of hair follicles on the scalp. The texture of the scalp appeared normal. He did not have finger clubbing and there was no other associated abnormality on physical examination.

Opthalmological and neurological examinations were within normal limits. A diagnosis of primary non essential cutis verticis gyrate was made.

Table 1: List of disorders associated with seconday CVG)

Pachydermoperiostosis (idopathic hypertrophic osteroarthropathy)
Roseenthal-Kloepfer syndrome
Endocrine
Acromegaly
Cretinism
Myxedema
Insulin resistance/acanthosis nigricans
Chromosomal
Turners syndrome
Klinefelter syndrome
Fragile X syndrome
Inflammatory
Eczema
Psoriasis
Paraneoplastic
Amyloidosis
Ehlers-Danlos syndrome

DISCUSSION

Cutis verticis gyrate (CVG) is a descriptive diagnosis for an abnormal-appearing overgrowth of the scalp in relation to the skull which may be congenital or acquired. The hypertrophy and associated folding of the scalp gives it s typical pattern[1]. The folds are usually anterior to posterior in nature and range from 2 – 12 folds. Although it was first used by Unna in 1907[2]. Garden and Robinson improved on the initial classification in 1984[3]. The condition may be classified into primary type of which there is the essential and the non essential form. There is also a secondary form.

The aetiology is unknown especially in the primary essential form. Most cases appear sporadic like our patient. However, autosomal recessive and dominant forms of inheritance

From: Department of Medical, University College Hospital, Ibadan, Oyo State.

Correspondence: **Dr. A. O. Ogunbiyi**, Department of Medical, University College Hospital, Ibadan, Oyo State.

have been described in some patients. In the primary non essential type, apart from an underlying genetic basis, it has been suggested that some endocrinological problem may be contributory. The primary non essential type occurs mainly in males after puberty and may disappear after castration. It has been suggested that this may be due to increase peripheral use of testosterone which was supported by a study in which male patients with CVG were found to have a reduced level of free testosterone compared with controls[4]. An associated with the fragile X syndrome and other fragile sites on chromosome [9, 10, and 12] and in a single case breaks at brands 3p 14 and 16q23, has been reported [5, 6]. The aetiology of CVG in the secondary forms is usually determined by the associated underlying disorder. The secondary type of CVG can occur at any age, and is usually asymmetrical. It may be congenital especially when nevi are involved and these are usually dermal. Tumors account for 25% of secondary CVG[7].

Patient with Turners syndrome usually develop CVG as a result of lymphoedema[8]. The prevalence of the different types of CVG is unknown in our environment. Reports else where shows it occurs in 0.5% of people with mental retardation in America [9]. In Sweden and Scotland it accounts for 0.71- 3.4% in mental homes [10,11]. Community studies suggested a prevalence of 1 in 100,000opulation for males and 0.0026 case in 100,000 for females. There is no racial predominance for this disorder.

The essential type is associated with neurological disorders such as seizures and microcephaly. It is also associated with psychiatric illness including schizophrenia and mental retardation. Opthalmological disorders, such as cataract, blindness and strabismus have also been associated with primary essential type [1,13,14]. The Primary non essential form is found in normal individuals like our patient and is quite rare [15] CVG can be secondary to various groups of disorders listen in (Table 1). Primary, non essential cuits verticis gyrata occurs more commonly in males. It has been documented in some families suggesting it may be genetic, although its mode of inheritance is uncertain. The folding of the scalp is usually noticed during adolescence. The folding starts gradually until it becomes static. There is usually no associated alopecia and the folds usually occur in the anterioposterior direction or may be horizontal especially when it involves the occipital area alone. In primary non essential CVG, the histology of the scalp is usually normal of there may be some evidence of hypertrophy of the adnexeal structure and increase in collagen deposits. In the secondary cases of CVG, the histology may be normal or suggestive of the underlying disorder especially if it is due to a nevus.

Management of patients with CVG includes classification into primary or secondary types. Neuroimaging studies especially computerized tomography (CT) scan and magnetic resonance imaging (MRI) should be performed on CVG presenting at birth or associated with mental retardation, neurological or opthalmological abnormalities. This helps to determine or exclude structural brain abnormalities. For primary types patients should be reassured. However for those opt for surgery, surgical reconstruction can be carried out either by simple excision or by tissue expansion. For secondary cases of CVG, the underlying cause should be treated. Surgical intervension by resection of tumours with wide margins are beneficial if there is an underlying nevus.

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