## **Case Report**

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# Cutis vertices gyrate: a case report

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#### **ABSTRACT**

Cutis verticis gyrata is a rare condition characterized by excessive growth of skin of the scalp, resulting in furrows and folds similar to gyri of the cortex of brain. It can be classified into two forms: primary (essential and non-essential) and secondary. We report a case of 18-year-old male patient with primary essential cutis verticis gyrata with rugosities over forehead and scalp and coarse facial features.

Keywords: Cutis vertices gyrata, Morphological aspects, Scalp

#### INTRODUCTION

Cutis vertices gyrata (CVG) is a descriptive term for a condition of the scalp manifesting as convoluted folds and furrows formed from thickened skin of scalp resembling cerebriform pattern. It may be classified into primary form (essential and nonessential) and secondary. The primary essential form is not associated with any other disorder except for the cutaneous alterations, whereas the primary non-essential type is associated with neurological and ophthalmological disorders. CVG may also develop secondary to local inflammatory or neoplastic processes of the scalp that cause hypertrophy of soft tissue of the scalp. The male to female ratio is 5-6 to 1

The presentation in most cases is before puberty. The lesions are mostly asymptomatic and typically affect the vertex and occipital regions of the scalp. The folds are usually symmetric in primary cutis verticis gyrata and asymmetric in secondary form. The condition can be diagnosed by observing the characteristic morphological aspects of the patient's scalp. The treatment consists of maintenance of local hygiene and surgical recession of the skin excesses.

Authors report a rare case of 16 years old male who presented with primary essential CVG.

#### **CASE REPORT**

Authors report a rare case of 16 years old male patient who presented to us with the chief complaint of asymptomatic rugosities over the scalp and forehead. These longitudinal folds were insidious in onset, initially localized over bilateral temporal areas which gradually progressed in size over 2 years to involve the entire scalp and extended over to involve forehead. Patient denied any similar family history, associated neurologic or psychiatric disorder, and consanguinity, usage of anabolic steroid drugs and past history of any scalp dermatosis.

Cosmetic appearance was the major concern for the consultation by the patient. Physical examination revealed multiple longitudinal spongy folds extending from frontal to occipital regions of scalp. These were symmetrical with no abnormality of hairs overlying the folds. There was associated coarsening of facial features with longitudinal furrowing and scarring over the forehead.

A complete laboratory workup was done which included CBC, LFT and RFT, Growth hormone levels, S. Testosterone levels and Thyroid profile.



Figure 1: Images of 16-YEAR-OLD male child with cutis verticis gyrate.

Radiological workup which included CT scan brain, x ray of skull bone was done to rule out any underlying neurological abnormality or secondary bone deformities.

Ophthalmologic and neurological evaluation was within normal limits.

#### DISCUSSION

Cutis vertices gyrate was originally described in 1837 by Jean-Louise- Marc Alibert, the term cutis vertices gyrate being proposed by Unna in 1907.<sup>1</sup>

CVG is classified into 2 forms Primary and Secondary. The Primary form can be divided in essential and non-essential.<sup>2</sup>

The primary essential form, presented by our patient, predominates in men (association 5 times more frequent compared to women). Usually, the clinical picture starts during or after puberty. There is no association with other comorbidities like neurological and ophthalmological diseases, and there is only formation of skin folds on scalp.<sup>3</sup>

The non-essential form presents association with several neurological manifestations (microcephaly and seizures, mental retardation, cerebral palsy, epilepsy) or ophthalmological changes (cataract, strabismus, blindness, retinitis pigmentosa).<sup>4</sup>

The secondary form may arise from use of drugs like anabolic steroids. It may also be associated with inflammatory and neoplastic processes that cause changes in scalp structure such as pituitary tumors, tuberous sclerosis, intracerebral aneurysm, amyloidosis, acne conglobate, syphilis, diabetes mellitus type 2.5 It may also

be associated with various syndromes like Turner syndrome, Noonan syndrome and fragile X syndrome.

Primary CVG presents as symmetric scalp folds which usually extend anteroposteriorly from vertex to occiput and can present transversally in the occipital region. The pathological examination may reveal an essentially normal skin structure or skin with thickened connective tissue with hypertrophy or hyperplasia of adnexal structures.

Differential diagnoses include acromegaly, cutis laxa, cylindroma and inflammatory diseases of the scalp8.

The treatment for primary essential CVG may be symptomatic or surgical. Orientation regarding local hygiene care is necessary to avoid secretion accumulation with unpleasant odour. Surgical treatment may be performed with the goal of improving the clinical aspect as well as cosmetic appearance.<sup>9</sup>

Surgical options include total resection of the lesion and grafting, placement of tissue expander in healthy skin and later grafting and partial resection of abundant portion of lesion.<sup>10</sup>

#### **CONCLUSION**

Primary essential CVG is an extremely rare condition where the primary essential form can be diagnosed after excluding neurological and ophthalmological conditions as well as other diseases that are possibly associated with onset of cutis vertices gyrata. It can be treated surgically with resection of thickened excess skin in coronal and sagittal axis.

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