

Case Report

A case of cutis verticis gyrata associated with epilepsy

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Abstract Cutis verticis gyrata (CVG) is characterized by thickening of the scalp, which becomes raised to form ridges and furrows resembling cerebral gyri that cannot be flattened by traction or pressure. The etiology is diverse, since different collections of cell types may be responsible for outward convoluted appearance. Herein a case of CVG associated with epilepsy is reported in a young girl in view of the clinical rarity of this association.

Key words

Cutis verticis gyrata, epilepsy, scalp, cerebral gyri.

Introduction

Cutis verticis gyrata (CVG) is a rare and slowly progressive deformity of the scalp with thick gyrated skin folds and ridges resembling gyri of the brain cortex. Those folds can lead to local skin infections, or a social and cosmetic complaint. CVG can be classified into two forms: primary (essential and non-essential) and secondary. The anatomy of the scalp and its attachment are such that folds and gyri are produced when cells infiltrate beneath the epidermis. The etiology is diverse, since different collections of cell types may be responsible for outward convoluted appearance, and range from inflammatory or hamartomatous infiltrations to neoplastic proliferations.¹

Case report

A 12-year-old girl presented with a history of

deep furrows and convolutions over scalp. These scalp corrugation had been there since birth and were progressively increasing in size. There was no history of any other skin, mucosal, soft tissue, or bone abnormality. She had no history of any inflammatory condition of skin or scalp. There was a history of epileptic seizures since childhood. Apart from seizures, her medical history was insignificant. She had no personal history of mental retardation, cerebral palsy, psychiatric disorder or ocular disorder. There was no family history of consanguinity, similar scalp condition or any bony deformity.

Cutaneous examination revealed multiple furrows and folds of 0.5-0.7 cm in depth and 4-5 cm in length, arranged anteroposteriorly along the scalp (**Figure 1**). There was no palmer hyperhidrosis, coarsening of facial features or clubbing. Systemic examination including ophthalmological examination was normal. Psychometry suggested a normal mental age and IQ. Routine hematological, biochemical and radiological investigations, including MRI brain were normal. EEG showed generalized paroxysmal epileptiform activity. Scalp biopsy was not done because her parents were unwilling

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Figure 1 Multiple furrows and folds of 0.5-0.7 cm in depth and 4-5 cm in length, arranged anteroposteriorly along the scalp.

for the same. Based on these clinical findings, a diagnosis of cutis verticis gyrata associated with epilepsy was entertained. The patient was referred to a neurologist for antiepileptic medication and advised plastic surgery for her scalp.

Discussion

Cutis verticis gyrata is a descriptive term for a condition of the scalp, in which deep furrows and convolutions are seen, that resembles the outer surface of the cerebrum. Though cutis verticis gyrata was coined by Unna in 1907 for cerebriform appearance of the skin, Fisher (1922) provided etiopathological details of the disorder. In general there are two main variables of this disease: primary and secondary. Primary type is further divided into essential and non-essential forms. The former may exist as a solitary finding, with no association with neurological and ophthalmological disease and the latter may be associated with mental

retardation, epilepsy and other brain or ophthalmologic abnormalities.^{2,3} In primary CVG, onset is after puberty, with a normal histology or revealing increased connective tissue or epidermal appendages. Secondary CVG may appear at any age, skin folds are usually asymmetric and histology reveals various alterations depending on underlying causes such as tumors, neurofibromas, cerebriform intradermal nevi and inflammatory conditions. Systemic disorders associated with secondary CVG include acromegaly, myxedema or amyloidosis.⁴

The underlying pathology responsible for thickening of the scalp lies in abnormal proliferation of collagen and reticular fibres, deposition of mucopolysaccharides in corium and epithelial hyperplasia.

CVG can also occur as a part of pachydermoperiostosis.⁵ This syndrome includes clubbing of the digits, periosteal new bone formation, coarsening of facial features, oiliness and furrowing of the skin of face, as well as that of scalp, seborrheic hyperplasia and hyperhidrosis of hands and feet.

A distinct Lennox-Gastaut syndrome has been described to show a symmetrical spike wave discharges at less than 3Hz even in sleep.⁶ Rotational traction over scalp hair can produce CVG in normal person.⁷

Treatment of CVG depends upon the etiology.⁸ Scalp reductive procedures have been performed for cosmetic reasons.

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