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Case Report

Rare Case of Cutis Vertex Gyrata

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Abstract
Cutis verticis gyrata is characterized by excessive formation of scalp skin. It may be primary (essential and nonessential) or secondary. In the primary essential form it presents only folding skin formation on the scalp, mimicking cerebral gyri, without associated comorbidities. We report a rare case of a 21 year-old male patient with primary essential cutis verticis gyrata.
Keywords: *Hypertrophy, Scalp dermatoses, cutis verticis gyrata*

Introduction

Cutis verticis gyrata (CVG) is a rare, benign deformity of the scalp characterized by excessive proliferation and hypertrophy of the skin and subcutaneous tissue. It can be classified as primary essential, primary non-essential, or secondary. Primary essential CVG occurs without any associated comorbidities, whereas primary non-essential CVG occurs with neurological or ophthalmological abnormalities. Secondary CVG usually occurs due to inflammatory or neoplastic changes in the scalp or in individuals with any underlying systemic illness.[1,2,3]

Case Report

A 21 years young male presented to us with asymptomatic thickening of the scalp skin with folds that were noticed incidentally . He did not have any scalp dermatoses in the past. There was no history of trauma, or any other known comorbidities. Family history was not significant. The physical examination revealed multiple, symmetrical, longitudinal folds and furrows over the frontal, parietal, occipital and vertex regions of his scalp. There was no associated thickening or furrowing of the face. Figure 1 showing preoperative images.

Neuropsychiatric and ophthalmological examinations were performed and found to be normal. Blood investigations complete blood count, liver function tests, renal function tests, fasting lipid profile, blood sugar, thyroid stimulating hormone were found to be normal. His hepatitis B and HIV tests were found to be

negative. Computed tomography of the skull cutaneous thickening with skin folding (fig 4). MRI brain revealed a thickened scalp with marked ridges and furrows of a gyriform appearance in the frontoparietooccipital region, suggestive of cutis verticis gyrata with no underlying defects (fig 3A & 3B).

Patient posted for surgery in supine position and fibroadhesiolysis was done in supragaleal plane and excision of excess scalp skin was done and wound closure done in layers. Figure 2 showing post operative images.

Histopathology was sent that reveals epidermal lining with very focal hyperplesia. The epidermis is showing dense irregular connective tissue and contains many accessory structures as skin appendages with muscle and blood vessels and nerve fibers. The hypodermis shown loose connective tissue with fibrofatty tissue and there is laying down of collagen however it is not dense. Section from galea shows increase in the connective tissue and encircling the adnexa. At places there is a thick layer of collagen.

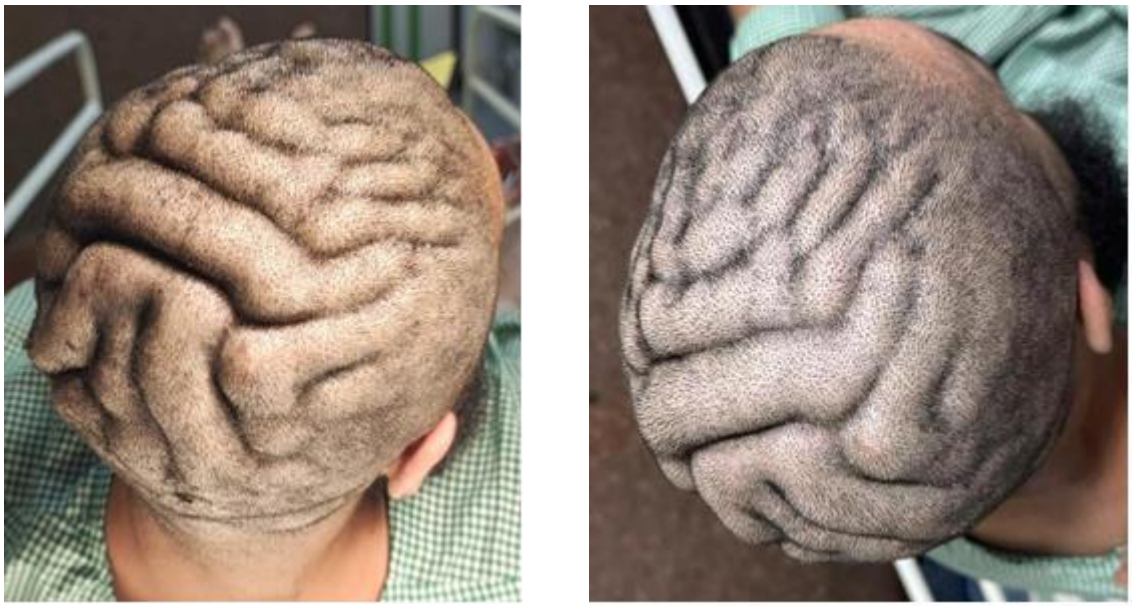

Figure: 1 Pre Operative)

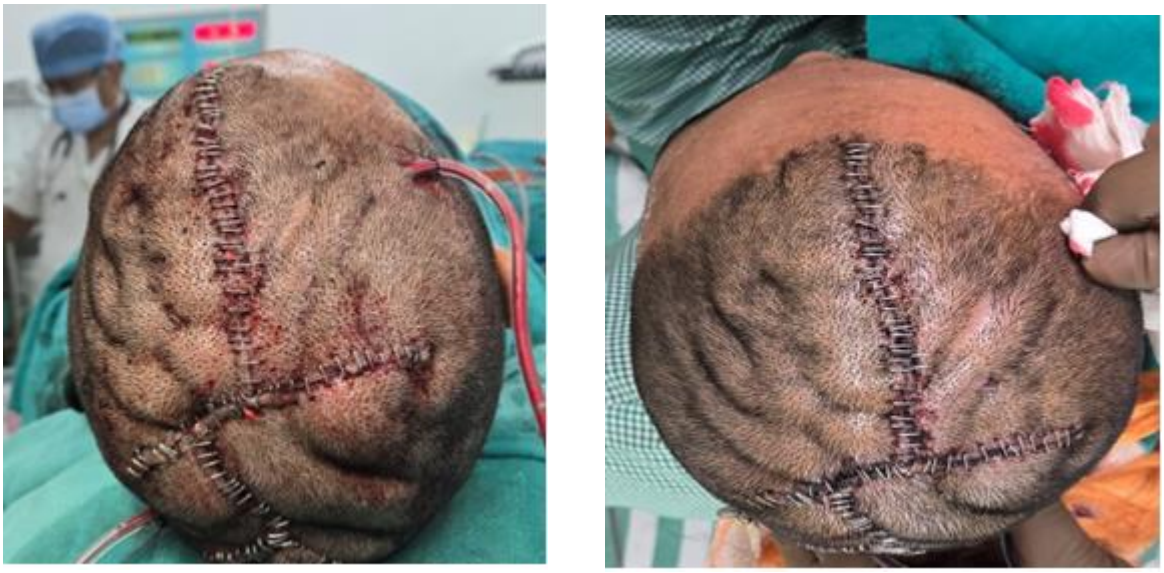

Figure: 2 Post Operative)

Discussion

CVG is characterized by excessive growth of the scalp skin and the formation of furrows that resemble a cerebriform pattern. The first case with a characteristic cerebriform appearance of the scalp was described by Alibert in 1837. In 1953, Polan and Butterworth classified CVG into two forms: primary and secondary.[4,5,6]

The estimated prevalence of CVG is 1 in 100,000 males and 0.026 in 100,000 females in the general population. Primary CVG is often noted during or after puberty. The male preponderance and postpubertal onset have been attributed to hormonal influences.[2] The exact reasons for the rarity of CVG among females are not known. Only three cases of primary CVG have been reported among females in the medical literature.

Primary CVG presents as symmetrical scalp folds, which usually extend anteroposteriorly from the vertex to the occiput. There may be around 2–30 folds, with varying widths (0.75–4 cm) and depths (a few millimeters to 1 cm).[6] CVG is often asymptomatic in most patients. At times, they can present with non-cosmetic issues such as headaches, itching, hair loss, dysesthesia, tenderness, local infections, maceration, or an unpleasant odor (fetid or butyric odor).[6]

The primary non-essential form of CVG presents with several neurological manifestations (microcephaly and seizures, mental retardation, schizophrenia, cerebral palsy, and epilepsy) or ophthalmological changes (cataract, strabismus, blindness, and retinitis pigmentosa). Around 0.2–4.5% of patients with a primary non-essential type of CVG have associated intellectual disabilities.[7]

Secondary CVG can occur due to various conditions like:

Dermatological causes: Eczema, Psoriasis, Folliculitis, Impetigo, Erysipelas, Pemphigus, Dariers, Pachydermoperiostosis, Acne conglobata, Cutaneous focal mucinosis, Dermatofibroma, Collagenoma, Neurofibroma, Cerebriform intradermal nevus, Fibromas, Cylindromas, Naevus lipomatosus, Connective tissue naevi

Malignancy: Pituitary tumours, Intracerebral aneurysm, Leukemia, T-cell lymphoma

Infections: Syphilis, Acquired immunodeficiency disease syndrome

Systemic disorders: Acromegaly, Tuberous sclerosis, Amyloidosis, Myxoedema, Acanthosis nigricans, Diabetes mellitus type 2

Syndromes: Noonan syndrome, Beare-Stevenson syndrome, Ehlers-Danlos Syndrome, Michelin tire baby syndrome, Apert syndrome, Turner syndrome, Fragile X syndrome Hyper IgE syndrome,

Drugs: Anabolic steroids

The differential diagnoses of CVG are pachydermoperiostosis, acromegaly, cutis laxa, cylindroma, cerebriform intradermal nevus, and inflammatory diseases of the scalp. Pachydermoperiostosis causes furrowing of the scalp as well as the face.[8] CVG can be differentiated from cutis laxa by giving traction and looking into whether folds can be flattened or not.[5]

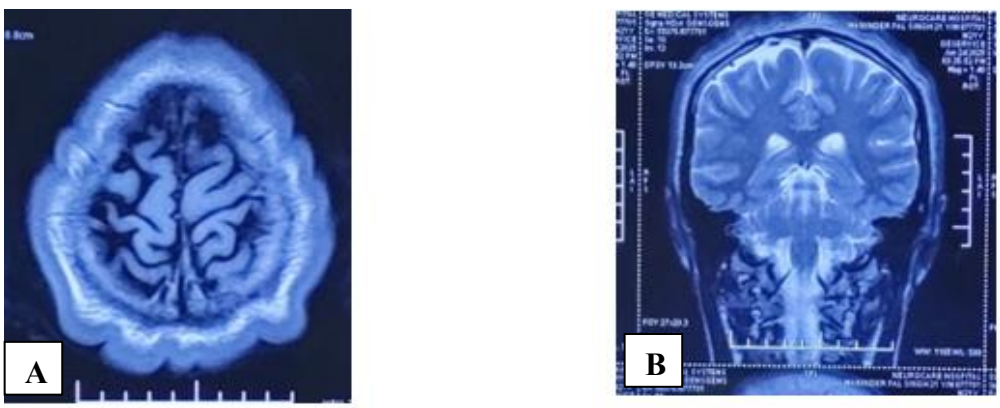

Figure: 3 Investigations to be performed in a case of cutis verticis gyrate

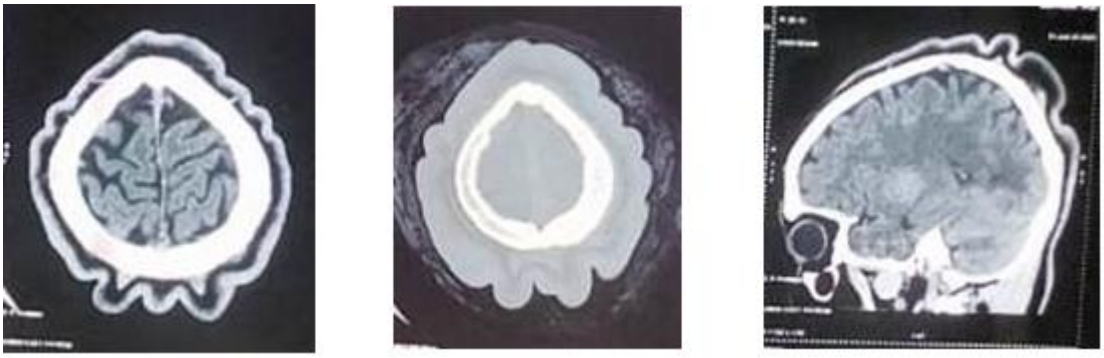

Figure: 4 Investigations to be performed in a case of cutis verticis gyrat

Laboratory investigations: Complete blood count, Liver and renal function tests, Basic metabolic profile, Total iron binding capacity, Blood sugar, Insulin, insulin-like growth factor-1, Thyroid function tests, Growth hormone level, Serum cortisol, Serum dihydrotestosterone, testosterone, Serum cholesterol, ANA, VDRL, HIV
Radiological investigations: Plain radiographs, Computed tomography, MRI brain.

A scalp biopsy is essential to exclude primary CVG from other causes of secondary CVG. Histopathology of primary essential CVG shows normal epidermis. The dermis may show either hyperplasia of the pilosebaceous unit and hair follicle, or an increase in the collagen bundles. In secondary CVG, histopathology depends on the underlying pathology.[3,6] Radiological investigations like X-ray, CT, and MRI help in ruling out secondary causes like intracranial and scalp tumors, intracerebral aneurysms, and pachydermoperiostosis.

The treatment for primary essential CVG includes symptomatic treatment such as maintenance of local hygiene, oral antihistaminics, topical steroids, and antibiotics in case of any secondary infections. Surgery is indicated only for cosmetic appeal, psychological concerns, and in case of any complications. For smaller folds, local excision and primary closure can be performed. The larger fold may require tissue expansion and staged excision or partial resection of the most abundant section of the lesion.[2] Management of the underlying cause is the mainstay of treatment in secondary CVG.

Conclusion

The occurrence of primary essential CVG among males is extremely rare. The diagnosis of primary essential CVG can only be made after ruling out the causes of primary non-essential and secondary causes of CVG. This rarity of disease warns more attentiom to diagnose the disease and to report the cases for further knowledge.

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