

An extremely rare case of primary cutis verticis gyrata: A comprehensive report in a female patient

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Abstract

Cutis Verticis Gyrata (CVG) is an unusual scalp condition characterized by excessive skin growth, resulting in prominent folds and furrows that resemble the cerebral gyri of the brain cortex. The diagnosis and classification of CVG are primarily based on clinical findings. Complementary investigations are essential to rule out any potential underlying systemic or local pathologies. In this study, we present a case of Primary Essential CVG that gradually expanded across the occipital region and significantly affected the patient cosmetically. Female presentation of CVG is extremely rare in the literature; therefore, this case provides valuable insight into the presentation and management of this disorder in female patients. The primary purpose of this case report is to describe the clinical features, diagnostic process, and surgical intervention for CVG, and to contribute to the literature.

Keywords: Cutis verticis gyrata, vertex, benign lesion

Introduction

Cutis Verticis Gyrata (CVG), also known as *pachydermia verticis gyrata* or *cutis verticis plicata*, is a rare scalp skin disorder that includes redundant scalp skin, leading to deep grooves and ridges that imitate cerebral convolutions [1]. This condition can result from congenital or acquired reasons. Histopathological microscopic findings typically demonstrate benign cutaneous hypertrophy. Redundancy of the scalp skin, forming thick skin folds and grooves, is also commonly observed among the macroscopic findings of this condition [2]. CVG is classified into three main categories: Primary Essential CVG, Primary Non-Essential CVG, and Secondary CVG. Primary Essential CVG occurs without associated systemic conditions, whereas Primary Non-Essential CVG can be linked to underlying neuropsychiatric or ophthalmologic disorders. Secondary CVG is mostly connected to systemic diseases or the use of medication [3]. Understanding the pathogenesis of CVG remains a challenge, and further research is needed to clarify its etiology and genetic implications.

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Informed Consent

The authors stated that the written consent was obtained from the patient presented with images in the study.

Conflict of Interest

No conflict of interest was declared by the authors.

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Case presentation

A 27-year-old female patient with no significant medical history presented with progressive scalp thickening and furrowing over three years. Her primary complaint was cosmetic; however, she also reported mild pruritus, maceration, and localized hair loss. There were no similar conditions in her family history. She denied consanguinity, anabolic steroid use, scalp infections, or systemic diseases. Neurological and ophthalmological examinations were unremarkable.

A punch biopsy was taken from the central zone of the scalp skin under local anesthesia. Histopathological evaluation revealed normal epidermal and dermal structures with no pathological alterations. Systemic assessments, including laboratory tests for thyroid function (TSH, free T4), complete blood count (CBC), fasting blood glucose, lipid profile, and syphilis serology, were within normal limits. Imaging studies, such as magnetic resonance imaging (MRI) and computed tomography (CT) scans, were conducted to exclude secondary causes related to structural anomalies or underlying malignancies, as well as to rule out conditions like acromegaly and pachydermoperiostosis. Skull and sella turcica CT and MRI showed no change. Serum growth hormone levels were within the normal range. On physical examination, the scalp exhibited soft, non-tender thickening with well-defined cerebriform folds predominantly affecting the parietal and occipital regions, covering an estimated 23 x 14 cm area. The furrows followed an anterior-to-posterior direction, with the central region demonstrating the greatest prominence. Manual traction failed to flatten the folds. Given these findings and the absence of any systemic condition, a diagnosis of Primary Essential CVG was established. A psychological assessment was conducted to evaluate the impact of the condition on the patient's mental well-being. It was observed that she had moderate distress associated with her altered appearance.

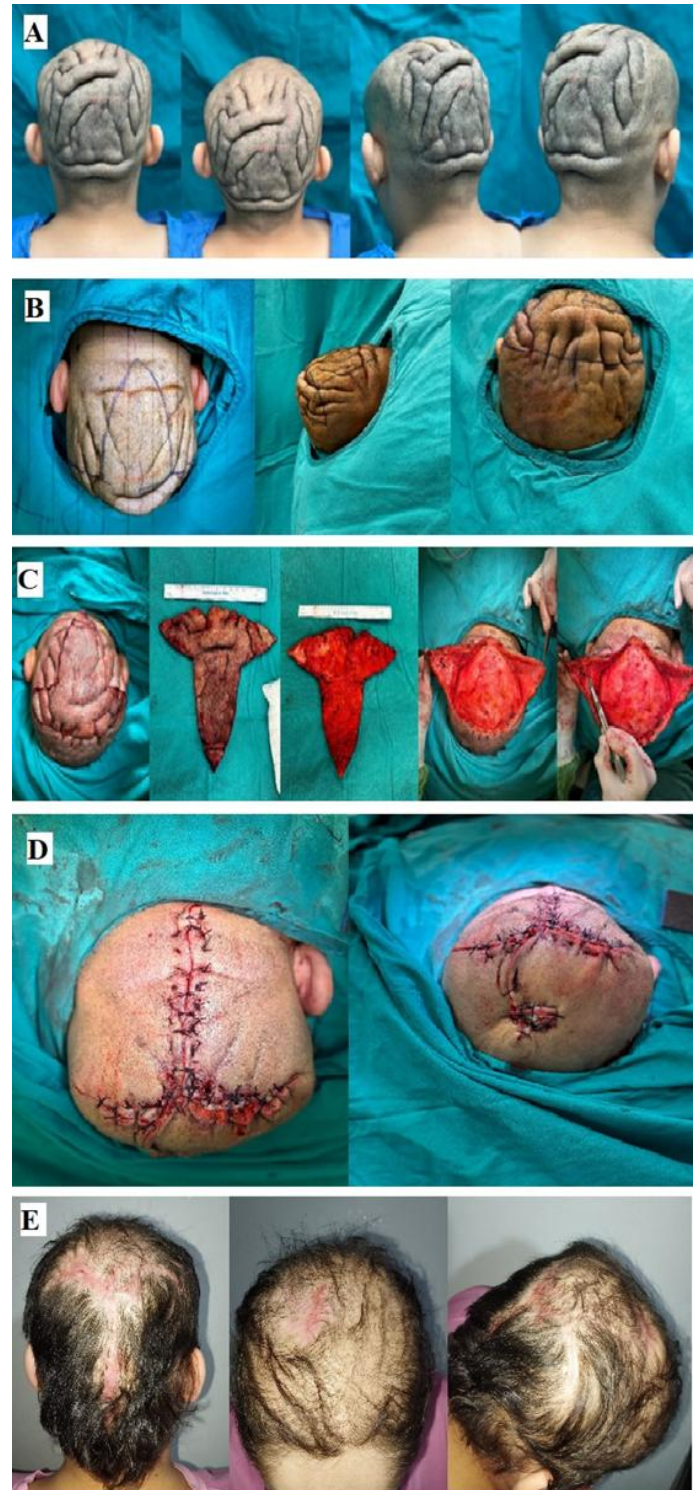
A written informed consent form was obtained from the patient for the use of her data and images in a scientific study.

Surgical Management

Considering the significant scalp deformity and the patient's aesthetic concerns, surgical intervention was planned. A total excision of the excessive scalp tissue was performed under general anesthesia with orotracheal intubation. The patient was positioned prone for optimal exposure of the occipital deformity. Following the skin tension assessment and excision marking, 0.4 mg epinephrine was infiltrated into the subgaleal and subcutaneous planes to minimize bleeding. A fleur-de-lis incision pattern was drawn, and tissue removal proceeded along the anteroposterior axis. Dissection was performed on the subgaleal plane and extended toward the parietal and occipital regions, taking care to preserve the occipital artery to maintain flap viability. Scoring techniques were applied to facilitate tension-free closure. Master sutures were placed centrally for primary closure. The final excised tissue measured 240 mm in length and 150 mm in width. A small rotational flap from the parietal region was utilized to assist in closure of the vertex defect. Closure was successfully achieved. Postoperative recovery was uneventful. The patient was closely monitored for potential complications such as wound dehiscence, infection, and hematoma formation.

Postoperative follow-up at one, three, and six months revealed a well-healed surgical site without recurrence of scalp furrowing. The patient expressed significant satisfaction with the cosmetic outcome and reported no discomfort or functional impairment. The complete preoperative and postoperative course is presented in Figure 1. Long-term monitoring will continue to assess any potential recurrence or other concerns.

Figure 1: The patient's progress at all stages before and after surgery (A: Pre-operative imaging after hair shaving, B: Planning of fleur-de-lis incision line, C: Incisions, excision, flap elevation, and scalp skin scoring, D: Immediate post-operative photo after skin closure, E: Post-operative 3-month result)



Discussion

CVG was first described by Jean-Louis-Marc Alibert in 1837. The term “*cutis verticis gyrata*”, which has been the accepted terminology ever since, was coined by Unna in 1907 [4]. Primary Essential CVG has no relation to any other abnormalities. Primary Non-Essential CVG may be associated with neuropsychiatric and ophthalmologic abnormalities [5]. Secondary CVG is mostly related to underlying diseases or causes such as inflammatory, neoplastic, metabolic conditions, and drug use (*i.e.*, growth hormone agonists) [6]. In particular, acromegaly and pachydermoperiosteosis (idiopathic hypertrophic osteoarthropathy) are disorders that often occur with secondary CVG. Turner syndrome, Klinefelter syndrome, and fragile X syndrome have been linked to primary non-essential CVG. The only known familial form of CVG is associated with primary pachydermoperiostosis [7].

Cutis verticis gyrata is predominantly seen in men, with a reported prevalence of approximately 1 in 100,000 males and 0.026 in 100,000 females [1]. A hormonal basis for CVG has been proposed due to its male predominance and postpubertal onset of this disorder. However, the potential role of genetic transmission and the pathophysiology of CVG remain ambiguous. Primary Non-Essential CVG usually occurs in postpubertal men. It has often been associated with intellectual disability or neuropsychiatric disorders such as seizures and schizophrenia. The folds of Primary Non-Essential CVG are usually symmetrical and run in an anteroposterior direction, involving the vertex and occiput [8]. Histopathological imaging may be normal or may show thickened connective tissue with hypertrophy or hyperplasia of adnexal structures [7]. Contrary to Primary CVG, Secondary CVG is relatively more common and can occur at any age. The skin folds of Secondary CVG are usually more asymmetrical than those of Primary CVG, and they may extend to the forehead region. The CVG form secondary to neoplasms tends to present as a localized area of scalp furrowing, which can then progress to the entire scalp [8].

In many cases reported in the literature, the patients are male, and the incidence in female patients has been reported to be very low. In male patients, the lesion is usually single and measures 5-8 x 3-7 cm. In contrast, the lesion in our case measured 24 x 15 cm and occurred in a female patient. Furthermore, our research did not reveal any secondary underlying cause or findings suggestive of primary non-essential CVG. This shows that our case is particularly rare and unique.

Conclusion

This case reports a rare occurrence of Primary Essential CVG in a young female patient. Surgical excision resulted in an effective outcome, with a smooth postoperative recovery. This report emphasizes the importance of ruling out secondary causes and underscores the role of surgical intervention as a viable treatment option for patients with symptomatic or cosmetically concerning CVG. Further research is needed to enhance understanding of the genetic and pathological basis of CVG and to develop more targeted management strategies. Future studies should focus on long-term recurrence rates and functional outcomes.

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