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Cutis verticis gyrata associated with drug-resistant epilepsy: A case report and review of the literature

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Abstract

Introduction. Cutis verticis gyrata is an uncommon benign dermatological disorder characterised by hypertrophic scalp folds resembling cerebral gyri. The primary form is subdivided into essential and non-essential; the latter associated with neurological abnormalities. Its relationship with epilepsy and neurodevelopmental impairment remains insufficiently recognised. We report a case of primary non-essential cutis verticis gyrata in a patient with drug-resistant epilepsy.

Presentation of the case. A 29-year-old male, born preterm with perinatal hypoxia, developed seizures from the second day of life. He subsequently presented impaired awareness seizures with automatisms and focal to bilateral tonic–clonic seizures, evolving into drug-resistant epilepsy despite multiple anti-seizure therapies. He exhibited global developmental delay, microcephaly, intellectual disability, and right hemiplegia. Brain magnetic resonance imaging demonstrated generalized atrophy and bilateral parieto-occipital encephalomalacia. During early adulthood, progressively deep longitudinal folds became clinically evident over the parietal and occipital scalp. After exclusion of secondary causes, primary non-essential cutis verticis gyrata was diagnosed.

Discussion and conclusions. The delayed dermatological manifestation relative to longstanding neurological disease underscores limited understanding of its natural history. Increased clinical awareness and further investigation of shared pathogenic mechanisms are warranted.

Keywords: Cutis Verticis Gyrata, Drug-Resistant Epilepsy, Seizure, Dermatology, Neurology, Microcephaly.

Cutis verticis gyrata asociada con epilepsia farmacorresistente: reporte de caso y revisión de la literatura

Resumen

Introducción. La cutis verticis gyrata es una dermatosis benigna poco frecuente caracterizada por pliegues hipertróficos del cuero cabelludo que simulan circunvoluciones cerebrales. La forma primaria se subdivide en esencial y no esencial, esta última asociada con alteraciones neurológicas. Su vínculo con epilepsia y trastornos del neurodesarrollo continúa siendo insuficientemente reconocido. Se presenta un caso de cutis verticis gyrata primaria no esencial en un paciente con epilepsia farmacorresistente.

Presentación del caso. Paciente masculino de 29 años, nacido pretérmino con antecedente de hipoxia perinatal, quien presentó crisis epilépticas desde el segundo día de vida. Evolucionó con crisis focales con alteración de la conciencia y crisis tónico-clónicas focales a bilaterales, configurando epilepsia farmacorresistente pese a múltiples esquemas terapéuticos. Presenta retraso global del neurodesarrollo, microcefalia, discapacidad intelectual y hemiplejía derecha. La resonancia magnética cerebral mostró atrofia generalizada y encefalomalacia parieto-occipital bilateral. En la adultez temprana se hicieron evidentes pliegues longitudinales progresivos en regiones parietal y occipital. Tras excluir causas secundarias, se estableció el diagnóstico de cutis verticis gyrata primaria no esencial.

Discusión y conclusiones. La manifestación cutánea tardía frente a la cronicidad neurológica evidencia vacíos en la comprensión de su historia natural y respalda la necesidad de mayor reconocimiento clínico e investigación fisiopatológica.

Palabras clave: cutis verticis gyrata, epilepsia farmacorresistente, crisis epiléptica, dermatología, neurología, microcefalia.

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Introduction

Cutis verticis gyrata is an infrequent and benign skin condition characterized by excessive proliferation and hypertrophy of the skin and subcutaneous tissue (1). It is classified as primary cutis verticis gyrata and secondary cutis verticis gyrata. Primary cutis verticis gyrata is diagnosed when secondary causes are excluded. It frequently appears in puberty, more frequently in men, and is characterized by the formation of symmetric antero-posterior folds (2). Secondary cutis verticis gyrata is diagnosed, when it occurs because of a systemic disease, as acromegaly, leukaemia, neuroendocrine tumours or other neoplasms, tuberous sclerosis, syphilis, and inflammatory diseases like Graves' disease, psoriasis, or eczema (3-7). Furthermore, primary cutis verticis gyrata is divided into essential and non-essential forms. Primary essential cutis verticis gyrata occurs isolated from other anomalies, while primary non-essential cutis verticis gyrata is associated with other neurological or ophthalmological abnormalities (1).

Here, a case of a male patient with primary non-essential cutis verticis gyrata associated with epilepsy and microcephaly is presented.

Case report

A 29-year-old male, born in Bogota, presents to an Epilepsy clinic. He is the second child of the couple, and the pregnancy was complicated because of preeclampsia, requiring preterm birth by caesarean section with a normal neonatal adaptation, but needing hospitalization because of neonatal hypoglycaemia and perinatal hypoxia. The patient has no family history of epilepsy, intellectual disability or microcephaly. Congenital infections were excluded. Global neurodevelopmental delay, microcephaly, and behavioural alterations become evident in the following years.

From two days old, the patient had seizures characterized by jerks with tonic upgaze, and later in life, he started to present impaired awareness seizures, joint with oral automatisms and fixed gaze, together with focal bilateral tonic clonic seizures. At present, the patient has daily seizures despite treatment with multiple anti-seizure medications: valproic acid (250 mg BID), brivaracetam (100 mg BID), lacosamide (200 mg TID), and clonazepam (2 mg BID), configuring a drug-resistant epilepsy. Precipitating factors for seizures included recurrent urinary tract infections and changes in medications associated with administrative difficulties for the delivery of the anti-seizure drugs. He also received treatment with levothyroxine because of hypothyroidism diagnosed at 25 years old.

Other anti-seizure medications that have been proven effective include levetiracetam 500 mg BID, which was stopped due to irritability and gastrointestinal side effects; topiramate 50 mg BID, which was stopped because of somnolence and gastrointestinal side effects; carbamazepine 400 mg BID, which was stopped due to leukopenia; and ketogenic diet, which was not continued because of poor adherence and low body weight.

On physical examination, deeply furrowed skin folds were palpable throughout the scalp (Figure 1) associated with microcephaly, spastic dysarthria, right hemiplegia, and dystonic posture of the right hand. Brain magnetic resonance showed generalized atrophy and confluent areas of bilateral cystic parieto-occipital encephalomalacia (Figure 2). The video electroencephalogram showed diffuse beta activity with anterior predominance without apparent posterior dominant activity, which is compatible with diffuse encephalic involvement. Now the patient is being evaluated by a surgery epilepsy group, in the quest of better control of epilepsy. As the skin changes have been asymptomatic and nonspecific, treatment for cutis verticis gyrate has been planned. Figure 3 shows a summary of the main events described in the case report.

Discussion

Cutis verticis gyrata is a rare, but benign, dermatological condition characterized by the progressive thickening of the skin and the connective tissue around the cranium, especially on the scalp, in rare cases, the forehead. This thickening gives the skin a resemblance to the sulci and gyri of the brain (8). It is more prevalent in males than females (9), especially at the onset of puberty. This epidemiological distribution has led to theories about the influence of hormonal pathways in the development of this condition, particularly the role of the growth hormone and testosterone in stimulating dermal fibroblasts, sebaceous glands, and hair follicles (10).

This condition was first described in medical literature by Alibert in 1837, and since then, it has been

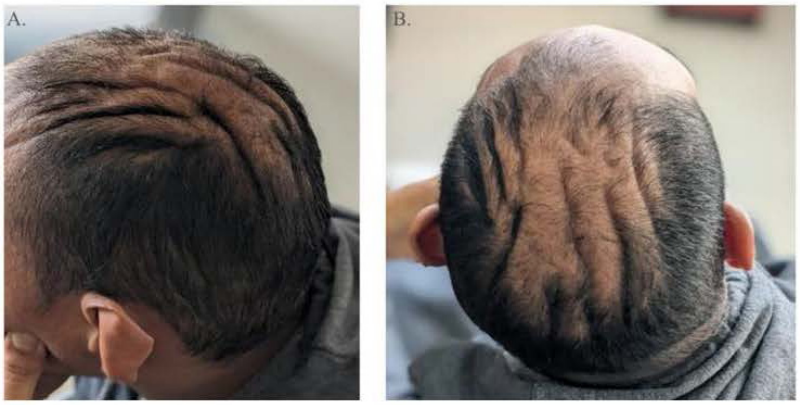

Figure 1. Clinical presentation of cutis verticis gyrata

Note. A and **B**. The clinical presentation of cutis verticis gyrata in a patient with drug-resistant epilepsy is characterized by longitudinal folds on the parietal and occipital scalp that resemble cerebral sulci.

Source: Patient's clinical history.

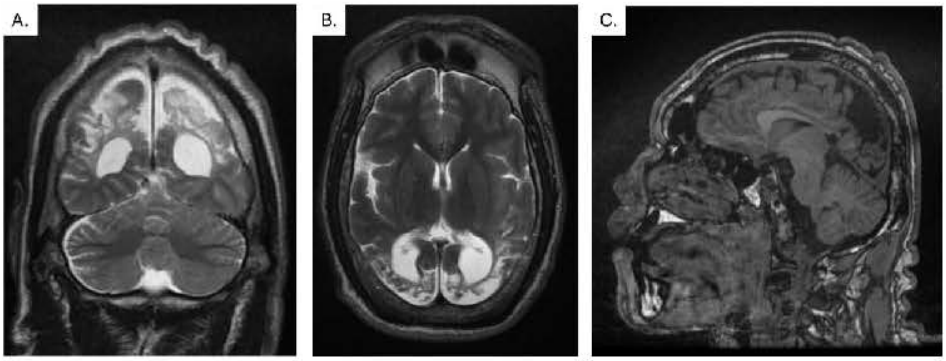

Figure 2. Brain magnetic resonance images

Note. A. T2 coronal sequence showing parieto-occipital atrophy, enlargement of lateral ventricles with associated ulegyria and cutis verticis gyrata in scalp. **B**. T2 axial sequence showing cranial hyperostosis, parieto-occipital atrophy, enlargement of lateral ventricles with associated ulegyria. **C.** T1 Fast Spoiled Gradient Recalled (SPGR) sagittal sequence showing right parieto-occipital atrophy with associated ulegyria and the gyri-like appearance in scalp (cutis verticis gyrata).

Source: Patient's clinical history.

referred to by various names, including paquidermia verticis gyrata, cutis vertices plicata, and even bulldog scalp syndrome (11). However, the most used and accepted term is the one employed throughout this report, which is also the term included in the MeSH database.

A case of primary non-essential cutis verticis gyrate is presented in a male patient diagnosed with drug-resistant epilepsy, microcephaly and intellectual disability. Although these diagnoses were made in the patient's early childhood, the dermatological changes to his scalp did not become clinically evident until later in early adulthood. Since then, they changes have increased in depth and spread from the occipital to the parietal region.

The association of the aforementioned neurological comorbidities with the later development of cutis verticis gyrata have not made things easier for the

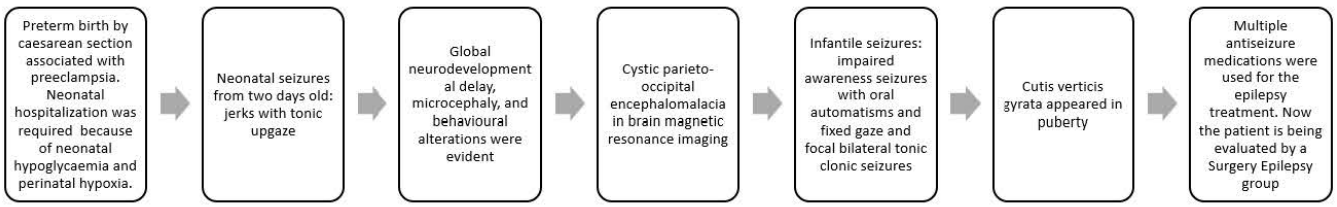

Figure 3. Timeline of the main events described in the case report

Source: Own elaboration.

patient and his support network, in words of his own mother: "the care burden is high due to severe functional and intellectual disability, along with the administrative obstacles for medication dispensing which lead to seizure aggravation and poor control of the epilepsy". She also mentions that, as his hair started to fall out in his late teens, the folds of his scalp became more evident, prompting indiscreet and unwanted questions and comments from some people. As if the expenses related to the epilepsy diagnosis were not enough, the apparition of cutis verticis gyrata led to some other economical responsibilities regarding special shampoos and moisturizer to avoid dryness.

Currently, the patient is under evaluation by an expert group of surgeons regarding his drug-resistant epilepsy with daily seizures. The cutis verticis gyrata diagnosis was made incidentally during the consultations regarding his epilepsy. Cutis verticis gyrata has been associated previously with drug-resistant epilepsy, intellectual disability, and microcephaly (12,13).

Although cutis verticis gyrata is a dermatological condition, it is frequently associated with intellectual disability and neurodevelopmental disorders. A 2016 case series of 62 patients with cutis verticis gyrata and intellectual disability described how cutis verticis gyrata presents as furrows separated by folds, which are usually symmetrical and arranged in parallel to each other in the anteroposterior direction (14). Other characteristic features described included a coarse, long face; prominent supraorbital ridges; a low narrow forehead; a high nasal bridge; a long nose; a broad jaw; a large and protruding ears and thick lips (14). Two brothers with microcephaly and hypothyroidism were reported, as the patient that was described.

Other cases of cutis verticis gyrata associated with epilepsy have recently been reported. Aldawari et al., described the case of a 15-year-old male with cutis verticis gyrata prior to the onsett of epilepsy. Magnetic resonance imaging revealed a subcortical white matter lesion affecting the right occipital lobe, accompanied by volume loss in the occipito-parietal lobe and posterior temporal region. This was pontentially linked to perinatal hypoxic insult, as the case of our patient (15). Another case report described in 2023 of a patient with epilepsy associated with cutis verticis gyrata and other brain malformations, including polymicrogyria, cortical dysplasia and hypoplasia of the corpus callosum (16). Cutis verticis gyrata has also been associated with Lennox-Gastaut Syndrome and neurological regression (17). It is also worth mentioning the case series in Colombia described by Hernández et al. They reported 65 male patients with cutis verticis gyrata, 46.15% had an intellectual disability, 43% had epilepsy and 27.7% had a language disorder. This data highlights the critical association between cutis verticis gyrata and neurological diseases (18).

Conclusions

This is a clear case of the primary non-essential form of the disease. Nonetheless, the late presentation compared with the long history of the patient's other pathologies, highlights the poor understanding in the medical literature of the disease's natural history. Despite its benign presentation and low incidence, it would be helpful to continue studying the underlying pathophysiological mechanisms that can explain primary forms of cutis verticis gyrata, especially those related to drug-resistant forms of epilepsy and cognitive impairments. This would enable preventive treatments to be offered to patients who

are at greater risk of developing this skin condition, which usually affects patients' emotional state due to its aesthetical impact.

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Ethical implications. Informed consent was obtained from the patient following the Nuremberg Code, ensuring privacy and confidentiality.

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References

1. Sankarapandian J, Palaniappan V, Karthikeyan K. Primary essential cutis verticis gyrata: A case report with a review of literature. Indian Dermatol Online J. 2024;15:313-5. https://doi.org/10.4103/idoj.idoj_210_23
2. García-Rodríguez I, Betancourt-Trujillo MR, Sánchez-Galván LA. Cutis verticis gyrata. Presentación de un caso y revisión de la literatura. Finlay. 2022; 2(3): 352-356.
3. Abu-Jamra F, Dimick DF. Cutis verticis gyrata. A report of a case secondary to acromegaly treated with plastic surgery and x-ray therapy. Am J Surg. 1966;111(2):274-7. https://doi.org/10.1016/0002-9610(66)90259-5
4. Rahman A, Mahmood A. Cutis verticis gyrata secondary to infiltrating ductal carcinoma breast. J Coll Physicians Surg Pak. 2012;22(2):120-2.
5. Bilen H, Atasoy M, Akcay G, Akcay M, Capoglu I, Gursan N, Akbas M. Elephantiasic pretibial myxedema and cutis verticis gyrata caused by Graves' disease. Thyroid. 2006;16(8):815-6. https://doi.org/10.1089/thy.2006.16.815
6. Passarini B, Neri I, Patrizi A, Masina M. Cutis verticis gyrata secondary to acute monoblastic leukemia. Acta Derm Venereol. 1993;73(2):148-9. https://doi.org/10.2340/0001555573148149
7. Moulias R, Noble JP, Auriol M, Ernst D, Bouvier AM, Devars Du Mayne JF, Congy F, Loeper J. Cutis verticis gyrata, hypertrophic gastritis, motrice diarhea, and horse's tall syndrome, a new association an apudoma (carcinoid or chemodectoma. Sem Hop. 1979;55(9-10):435-41.
8. Adôrno IF, Santos RFT, Nunes TF, Sandim GB, Marchiori E. Primary essential cutis verticis gyrata. Radiol Bras. 2019;52(4):276-277. https://doi.org/10.1590/0100-3984.2017.0218
9. Tucci A, Pezzani L, Scuvera G, Ronzoni L, Scola E, Esposito S, Milani D. Is cutis verticis Gyrata-Intellectual Disability syndrome an underdiagnosed condition? A case report and review of 62 cases. Am J Med Genet A. 2017;173(3):638-646. https://doi.org/10.1002/ajmg.a.38054
10. Rattagan M, De Francesco M, Kriebaum A, Ferraro F, Major C, Sharma D, Ojeda A, Martinez O, Musto AE. Cutis verticis gyrata: Two cases associated with drug-resistant epilepsy. Clin Case Rep. 2020;8(8):1365-1368. https://doi.org/10.1002/ccr3.2814
11. Shareef S, Horowitz D, Kaliyadan F. Cutis Verticis Gyrata. 2023 Jul 10. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 Jan–. PMID: 30969634.
12. Striano S, Ruosi P, Guzzetta V, Perone L, Manto A, Cirillo S. Cutis verticis gyrata--mental deficiency syndrome: a patient with drug-resistant epilepsy and polymicrogyria. Epilepsia. 1996;37(3):284-6. https://doi.org/10.1111/j.1528-1157.1996.tb00027.x
13. Chang GY. Cutis verticis gyrata, underrecognized neurocutaneous syndrome. Neurology.1996;47(2):573-5. https://doi.org/10.1212/WNL.47.2.573

14. Tucci A, Pezzani L, Scuvera G, Ronzoni L, Scola E, Esposito S, Milani D. Is cutis verticis Gyrata–Intellectual Disability syndrome an underdiagnosed condition? A case report and review of 62 cases. Am J Med Genet A. 2017;173(3):638–646. https://doi.org/10.1002/ajmg.a.38054

15. Aldawsari S, Aljughayman M, Alhamed A, Alhazza M, Almulhim N, Alnaim M, Siddiqu F. Cutis verticis gyrata and epilepsy, is there a typical patient?: A case report. Electron J Gen Med. 2025;22(2):638. https://doi.org/10.29333/ejgm/16010

16. Elhadri K, Raboua M, Amenzouy F, Boutaqiout B, Idrissi–Ouali N, Cherif Idrissi Ganouni N. A Case Report of a Cutis Verticis Gyrata–Intellectual Disability. Sch J Med Case Rep. 2023;11(5)825–827. https://doi.org/10.36347/sjmcr.2023.v11i05.020

17. Filosto M, Tonin P, Vattemi G, Bongiovanni LG, Rizzuto N, Tomelleri G. Cutis verticis gyrata, mental retardation and Lennox–Gastaut syndrome: a case report. Neurol Sci. 2001;22(3):253–256. https://doi.org/10.1007/s100720100022

18. Hernández JF, Espejo–Jiménez LM. Correa–Díaz RL, de la Espriella R. Cutis verticis gyrata en población masculina institucionalizada con enfermedad mental crónica en Colombia. Serie de casos. Acta neurol. Colomb. 2015; 31(4): 404–411. https://doi.org/10.22379/2422402259