A case of cutis verticis gyrata developing in a patient with primary scarring alopecia: A unique presentation of a rare disorder

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Key words: CCCA; central centrifugal cicatricial alopecia; cicatricial alopecia; cutis laxa; cutis verticis gyrata; cutis verticis gyrata-intellectual disability; CVG; CVG-ID; erythema; folliculitis decalvans; linear morphea; perifollicular scaling; primary essential; pruritus; scalp tenderness; scarring alopecia; secondary CVG; undulations; vertex hair loss.

REPORT OF CASE

A 39-year-old female with a medical history including hydronephrosis, kidney stones, hyperlipidemia, anxiety, and gastroesophageal reflux disease, presented to the dermatology clinic with complaints of scalp tenderness and pruritus with associated vertex hair loss. The patient's hair loss began 13 years prior, and she subsequently developed scalp undulations a few years later, which progressed in prominence over time. Scalp pruritus and tenderness within these undulations developed 3 years prior to presentation to our clinic. She was of normal intelligence; denied any history of ocular disease, scalp injuries, endocrine disorders, or tumors. The patient's current medications include fluticasone propionate, cetirizine HCl, and semaglutide. She denied a family history of autoimmune diseases and hair loss was experienced by her father only. On examination she was found to have 2 adjacent linear deep furrows running anterior to posterior, forming a single undulation from the midscalp to the right crown, traversing the area of hair sparsity. These undulations were palpable to touch and visible on magnetic resonance imaging of the scalp (Fig 1). On trichoscopic examination, this area also exhibited evidence of follicular dropout, perifollicular scaling, and erythema (Fig 2), with additional scalp bogginess in the surrounding area. Differential diagnoses were cutis verticis gyrata (CVG), linear morphea, and cutis laxa, as well as a scarring alopecia.

Abbreviations used:

AKN: acne keloidalis nuchae
CVG: cutis verticis gyrata

A 4-mm punch biopsy from the right crown revealed a gently papillated and hyperplastic epidermis with multiple dilated and compounded follicular infundibula opening up into the surface, which is suggestive of CVG (Fig 3). Additionally, polytrichia with perifollicular fibrosis was present most consistent with a cicatricial alopecia (Fig 4). The histologic differential included lichen planopilaris, central centrifugal cicatricial alopecia, as well as folliculitis decalvans.

The patient was treated with topical clobetasol 0.05% 5 times a week as well as intralesional triamcinolone 5 mg/mL, 1 mL, repeated every 1-3

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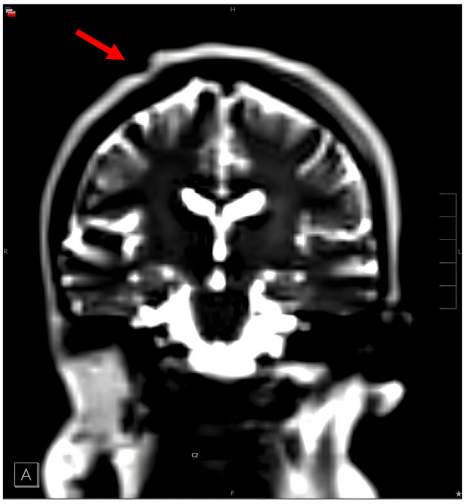

Fig 1. MRI Scout Imaging: *Red arrow* indicates the most prominent scalp undulation. *MRI*, Magnetic resonance imaging.

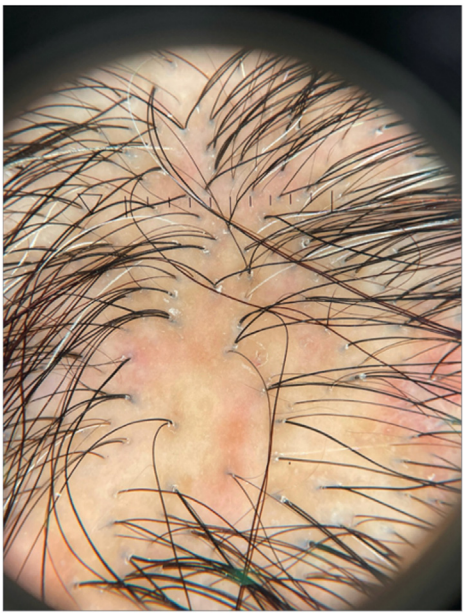

Fig 2. Trichoscopy/Dermoscopy findings of follicular dropout, perifollicular scaling, and erythema.

months. The patient was also started on oral minoxidil 0.625 mg daily for 1 month and increased to 1.25 mg daily thereafter. In addition to the scalp biopsy, other interventions included a chest x-ray to rule out lung cancer, a complete blood count, thyroid stimulation hormone levels, and thyroid peroxidase antibody. All of the results were normal.

DISCUSSION

CVG is primarily observed in men, with a prevalence of approximately 1 in 100,000 males and 0.026

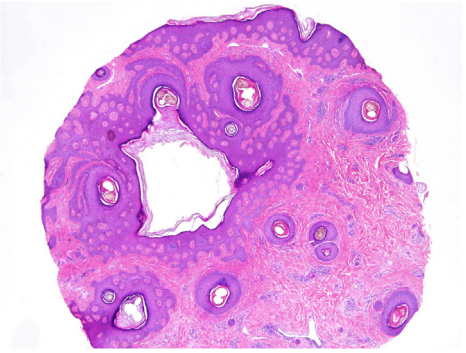

Fig 3. 4× Histology: Epidermis demonstrating multiple dilated and compounded follicular infundibula opening up to the surface.

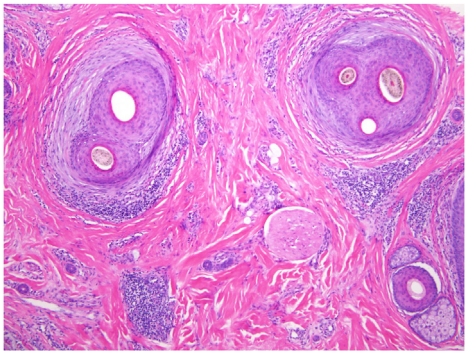

Fig 4. 10× Histology: Follicular units show fusion of hair follicles (follicular bigeminy and trigeminy) with prominent perifollicular fibrosis and a surrounding infiltrate characterized by lymphohistiocytic infiltrate with scattered neutrophils.

in 100,000 females.[1] The pathogenesis of CVG remains uncertain, though a hormonal etiology has been postulated due to the male predominance and postpubertal onset of the disorder.[1] The various presentations of the 3 categories of CVG are summarized in Table I. CVG-Intellectual Disability and secondary CVG have been linked to ophthalmological abnormalities, neurologic disease, and mental deficiency.[1] Secondary CVG is caused by underlying systemic inflammatory processes such as eczema and psoriasis. Primary scarring alopecias are rarely reported in association with CVG. There is 1 report of a patient with CVG, folliculitis decalvans, and acne keloidalis nuchae (AKN).[3] The authors note all 3 conditions are associated with dermal collagen thickening which may result from fibroblast growth factor activation. A database study conducted over 11 years from July 2009 to November 2020 involving a total of 108 patients with AKN at a dermatology clinic in Los Angeles revealed a higher incidence of CVG in patients with AKN. This study reported a

Table I. Diagnostic findings of CVG and cicatricial alopecia

	Epidemiology/clinical history	Physical examination	Trichoscopy/ dermoscopy	Pathologic features
CVG- Primary essential CVG[1]	Men > Women Postpubertal men before the age of 30 Idiopathic and typically no associated abnormalities	Symmetric, redundant scalp folds that exhibit deep furrows and convolutions, and the appearance mimicking that of cerebral gyri, furrows are typically anterior to posterior	Variable	Range from normal skin architecture to thickened connective tissue with hypertrophy or hyperplasia of adnexal structures with or without an associated increase in collagen fibers. The folds in secondary CVG are that of the underlying disease process. Epidermal hypertrophy and hyperplasia
CVG- Intellectual disability (CVG-ID)[1]	Usually associated with neurological and ophthalmological disorders	Folds may be more asymmetrical		
Secondary CVG[1]	May be associated with hormonal and inflammatory disease processes			Characteristics of underlying disease process
Primary Cicatricial alopecia[2]	Prevalence of approximately 7% CCCA and FD have higher prevalence in those of African descent CCCA more common in women FD more common in men	Well-circumscribed, areas of hair loss on the scalp. The hair loss may be accompanied by itching, burning, and pain. The affected area may also have a shiny, smooth appearance.	Broken hairs, perifollicular erythema, and white scales Peritubular hyperkeratosis	Perifollicular fibrosis, destruction of the hair follicle, dilated eccrine glands, and a lymphocytic or neutrophilic infiltrate.

CCCA, Central centrifugal cicatricial alopecia; *CVG*, cutis verticis gyrata; *FD*, folliculitis decalvans.

CVG incidence rate of 3.7% amongst patients diagnosed with AKN, which is significantly higher than the 0.001% incidence rate observed in the general population.[4] Another report is of a patient with scarring alopecia and pachydermoperiostosis, which is associated with CVG although the patient did not exhibit CVG.[5] There is a case of CVG concurrent with alopecia areata.[6] The authors postulate a possible genetic association as there is evidence of a mutation in the FGFR2 gene which may be involved in the development of CVG and this gene is located on a chromosome that has been linked with alopecia areata.

An increased understanding of the pathogenesis of CVG is necessary to determine possible associations with alopecia, including primary scarring alopecia.

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Conflicts of interest

Dr Shapiro is a consultant for Aclaris Therapeutics, Incyte, and Replicel Life Sciences. Drs Shapiro and Lo Sicco have been investigators for Regen Lab and are investigators for Pfizer. Dr Lo Sicco is a consultant for Pfizer and Aquis. MGB, LA, EK, CO, RHK, and EAR have no conflicts to disclose.

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