

A Unique Overlap of Scarring and Non-Scarring Alopecia in Primary Cutis Verticis Gyrata

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Abstract

Cutis verticis gyrata (CVG) is a rare scalp disorder characterized by cerebriform thickening and folding of the scalp skin. It can present as a primary essential form without underlying systemic involvement. Alopecia, classified as scarring or non-scarring, may rarely accompany CVG. We report a 55-year-old female with primary essential CVG exhibiting both non-scarring alopecia areata and scarring lichen planopilaris, which were confirmed histopathologically. No neurological, ophthalmological, or systemic abnormalities were present. Magnetic resonance imaging revealed no intracranial or cranial bone pathology. To our knowledge, this is the first reported case of simultaneous scarring and non-scarring alopecia in a patient with primary essential CVG. This rare coexistence may reflect the presence of multiple autoimmune pathways targeting different segments of the hair follicle. Clinicians should consider combined pathological mechanisms in unusual alopecia presentations associated with CVG.

Keywords: Cutis verticis gyrata, alopecia areata, lichen planopilaris, scarring alopecia, autoimmune overlap

INTRODUCTION

Cutis verticis gyrata (CVG) is a rare dermatological condition characterized by excessive thickening and folding of the scalp skin, resulting in a cerebriform appearance that mimics the surface of the brain. CVG is classified into three subtypes: primary essential, primary non-essential (associated with neuropsychiatric or ophthalmological disorders), and secondary (due to local or systemic causes such as tumors, inflammatory dermatoses, or trauma).¹ The pathogenesis of primary CVG remains poorly understood. Proposed mechanisms include hormonal imbalances, such as altered androgen metabolism, and abnormalities in connective tissue development. Histopathological findings in primary CVG are often non-specific, ranging from normal skin to hypertrophy of adnexal structures and increased dermal collagen. Alopecia is broadly divided into non-scarring and scarring types, depending on whether permanent destruction of hair follicles occurs. Alopecia areata (AA) is a common autoimmune form

of non-scarring alopecia, whereas lichen planopilaris (LPP) represents a lymphocytic scarring alopecia.² To date, only a few cases have been reported in the literature that associate alopecia with CVG, and none have described the coexistence of both scarring and non-scarring types in the same patient with primary essential CVG. This report presents a unique case illustrating this rare overlap.

CASE REPORT

A 55-year-old woman presented with progressive scalp deformity and hair loss. Ten years earlier, she had been diagnosed with seborrheic dermatitis and CVG based on scalp biopsy, but no follow-up was performed. On examination, alopecic patches and deep cerebriform folds were observed over the vertex and occipital scalp. Histopathology from these areas showed features of both scarring and non-scarring

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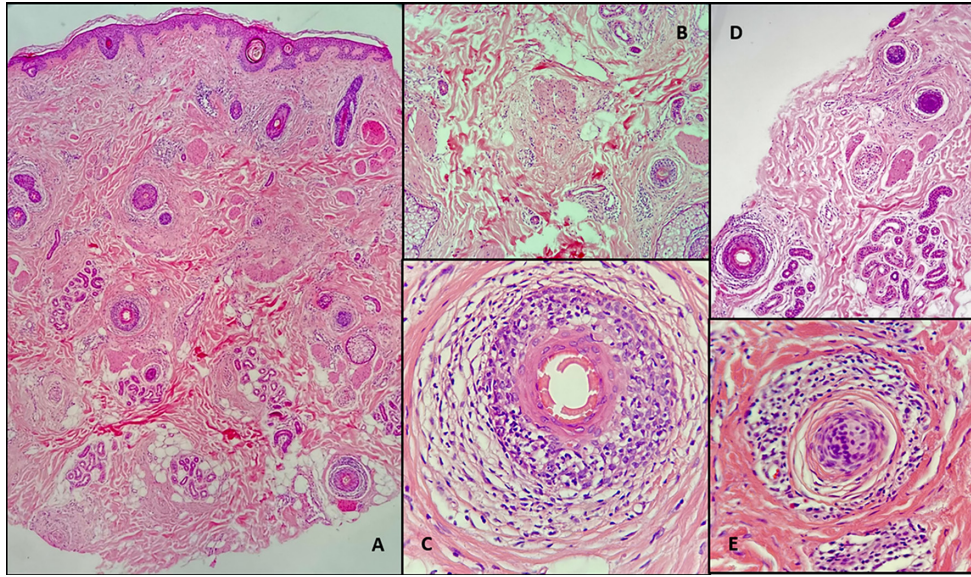


Figure 1. Histopathologic sections from scalp biopsies: (A) Follicular hypertrophy in dermis (H&E x 40). (B) Follicular scar tissue (H&E x 100). (C) Perifollicular lichenoid infiltrate at infundibular level, suggestive of LPP (H&E x 200). (D) Mixed pattern: LPP below, AA above in same field (H&E x 100). (E) Lymphocytic infiltration around anagen hair bulb (H&E x 200).
H&E: Hematoxylin and eosin, LPP: Lichen planopilaris, AA: Alopecia areata



Figure 2. The clinical image of the patient's scalp shows the CVG pattern in the occipital region
CVG: *Cutis verticis gyrata*

alopecia: perifollicular lichenoid inflammation and fibrosis consistent with LPP, along with peribulbar lymphocytic infiltrates characteristic of alopecia areata (Figure 1). Systemic evaluation, including cranial magnetic resonance imaging, revealed no neurologic or ocular involvement or intracranial pathology (Figure 2). She was started on topical corticosteroids and scheduled for regular dermatologic follow-up. Written informed consent was obtained from the patient for publication.

Discussion

CVG is an uncommon condition involving hypertrophy of the scalp skin, forming undulating folds that resemble cerebral gyri. It may present as a primary essential form, with no systemic associations, or secondary to underlying local or systemic conditions.^{1,3} Histopathology in primary CVG is often non-specific, though thickening of dermal collagen or adnexal hypertrophy may be observed. In our case, histologic features of CVG coexisted with findings diagnostic for both LPP (scarring alopecia) and AA (non-scarring alopecia).

Table 1. Cases of CVG with alopecia

Literature	Age	Gender	CVG type	Alopecia type
Buontempo et al. ⁷	39	F	Primary essential	Primary scarring alopecia
Anansiripun and Suchonwanit ⁸	24	M	Secondary	Scarring alopecia
Alonso Pereira et al. ⁹	29	M	Secondary	Folliculitis decalvans, folliculitis keloidalis nuchae
Bonalumi Filho et al. ¹⁰	43	F	Secondary	N/A
Alcántara González et al. ¹¹	48	M	Secondary	N/A
Yoo et al. ⁶	28	M	Primary essential	Alopecia areata
Mishra et al. ¹²	18	F	Primer essential	N/A
Saoji et al. ¹³	48	M	Secondary	N/A
Pai and Rao ¹⁴	25	M	Secondary	Scarring alopecia
Fox et al. ¹⁵	Newborn	F	Congenital	N/A
van Geest et al. ¹⁶	46	M	Secondary	N/A
Jeanfils et al. ¹⁷	43	M	Secondary	N/A
Hamm and Argent ¹⁸	41	F	Secondary	N/A
Yazici et al. ¹⁹	7	F	Secondary	N/A
Kanwar et al. ²⁰	25	F	N/A	Tractional

N/A: No answer, F: Female, M: Male, CVG: Cutis verticis gyrata

Although both LPP and AA are considered autoimmune disorders, they typically exhibit distinct immunopathological profiles. The co-occurrence of scarring and non-scarring alopecia in the same patient raises the possibility of a shared underlying autoimmune predisposition, potentially targeting different components or layers of the hair follicle unit. This aligns with theories proposing that variable follicular immune privilege collapse may contribute to divergent clinical outcomes.^{4,5} A literature review identified 15 prior cases of CVG with alopecia. Of these, one involved AA⁶ and one primary CVG with scarring alopecia (Table 1).⁷ However, to our knowledge, this is the first reported case of both forms of alopecia occurring simultaneously in a patient with primary essential CVG. Further studies are needed to clarify whether this dual pattern reflects coincidental findings or a broader, multi-level immune dysregulation affecting follicular structures in CVG patients.

Ethics

Informed Consent: Written informed consent was obtained from the patient for publication.

Footnotes

Authorship Contributions

Design: S.Y., Analysis or Interpretation: Ş.B., Literature Search: B.M.D., Writing: B.M.D.

Conflict of Interest: The authors declared that they have no conflict of interest.

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