Occipital alopecia in a young man

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Abstract

Lipedematous alopecia is a rare, non-androgenic form of alopecia that is challenging to diagnose, often requiring clinical-pathological correlation. The condition has been reported predominantly in African-American females, but more recently has been described in a broader demographic [1,2]. We describe a rare case of a young Caucasian man with isolated lipedematous alopecia who presented with a boggy, erythematous plaque with alopecia of the occipital scalp and subcutaneous thickening with lymphocytic dermal infiltrate and decreased anagen hairs on histology.

Keywords: lipedematous alopecia, non-androgenic alopecia

Introduction

Lipedematous alopecia is a rare non-androgenic form of alopecia reported predominantly in African-American females. Lipedematous alopecia is challenging to diagnose, often requiring attentive clinical-pathological correlation. More recently, lipedematous alopecia has been described in a broader demographic, including in Caucasians, Hispanics, and Asians [1,2]. Cases of lipedematous alopecia have been described in association with more common forms of alopecia, including androgenic alopecia, as well as in the absence of other types of alopecia [1,3]. Herein, we describe a case of isolated lipedematous alopecia in a young, adult, Caucasian male.

Case Synopsis

A young healthy man in his 20s presented to clinic with a single patch of alopecia overlying the occipital scalp. He reported gradual onset over the preceding 18 months without preceding trauma. He denied associated symptoms and experienced no improvement following a course of topical betamethasone valerate. Physical examination revealed a 5cm×3cm, poorly demarcated, erythematous, boggy plaque with overlying alopecia of the superior occipital scalp. The plaque was non-tender to palpation and without pustules or sinus tract formation (**Figure 1**). Complete blood count with differential, basic metabolic panel, and thyroid stimulating hormone were within normal limits. Punch biopsy and incisional biopsy of the lesion were performed.

A 4mm punch biopsy of the occipital scalp exhibited non-scarring alopecia with minimal lymphocytic inflammation and lipoatrophic changes of the subcutaneous tissue (**Figure 2A**, **B)**. Subsequent incisional biopsy demonstrated reduced density of terminal hair follicles with small adipocytes associated with mildly increased vasculature and fibrous tissue (**Figure 2C**, **D)**. Periodic acid-Schiff-diastase staining was negative for fungus and staining with colloidal iron revealed subtle collections of mucin in the dermis and subcutaneous fat. Given this constellation of findings, the patient was diagnosed with lipedematous alopecia.

Case Discussion

This case highlights the challenge of diagnosing lipedematous alopecia and the need for clinical pathological correlation in these instances when neither clinical nor pathologic findings are specific. Lipedematous alopecia is characterized by thickening of the scalp owing to expansion of the underlying subcutaneous fat with secondary

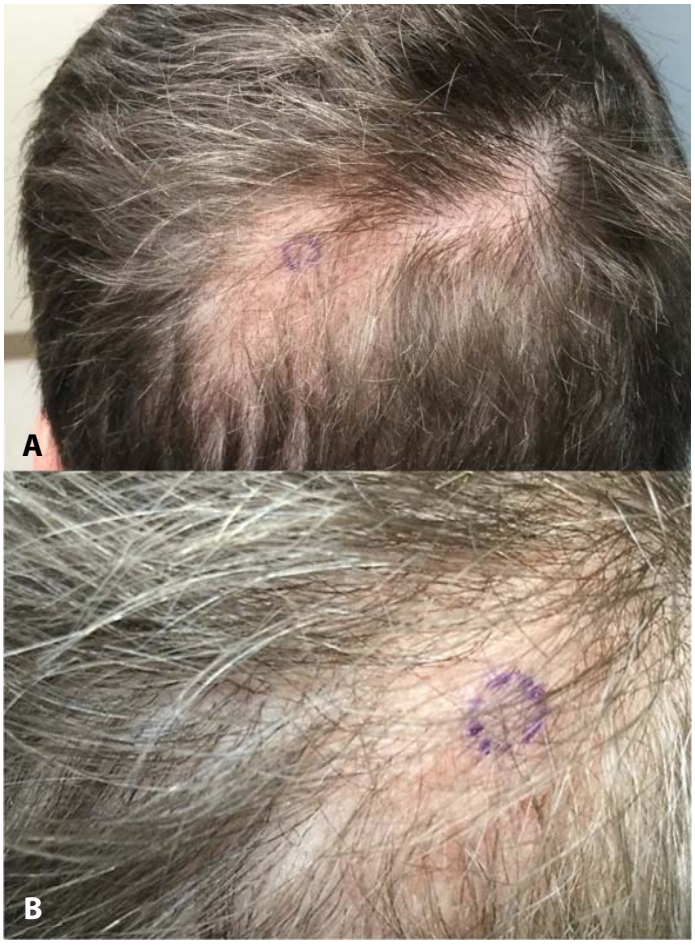

Figure 1. ***Lipedematous alopecia of the superior occipital scalp with a*** *5cm×3cm, poorly demarcated, erythematous, boggy plaque, shown in* ***A)*** *standard view and* ***B)*** *magnified view.*

alopecia, which can be focal or diffuse [1,3-5]. The characteristic palpable, boggy scalp is likened to "cotton batting" on gross examination and is a key to the underlying diagnosis [4]. The etiology and pathogenesis of this disease remains unknown, however, lymphatic dysfunction has been postulated as a primary contributing factor [3]. Leptin may play a role in the pathogenesis of lipedematous alopecia given that this hormone is involved in feedback mechanisms that regulate fat mass and distribution [6]. Another proposed theory involves metaplasia and displacement of adipose tissue [6].

Importantly, full thickness biopsy is necessary to confirm the histologic diagnosis and lipedematous alopecia will be missed in more superficial specimens as the pathological process is demonstrated in the subcutaneous tissue, whereas the epidermis and dermis appear largely unremarkable [4]. The subcutaneous architecture demonstrates thickened, edematous fat lobules and collagenous septae that have been observed to be disorganized in some cases [1,4]. As seen in our patient, deposition of mucin is variably present [1,3-5]. Hairs are typically normal and present in the appropriate ratio of anagen to telogen phase, although their associated bulbs may be atrophied or decreased in number [3,4]. A mild perivascular or perifollicular lymphoid infiltrate may be present [3-5]. There is discussion that the alopecia noted in lipedematous alopecia may be induced by elevated tissue oncotic pressure on hair bulbs [4].

Treatment of lipedematous alopecia is challenging with generally poor response to intra-lesional or topical corticosteroids [5]. Other treatments that

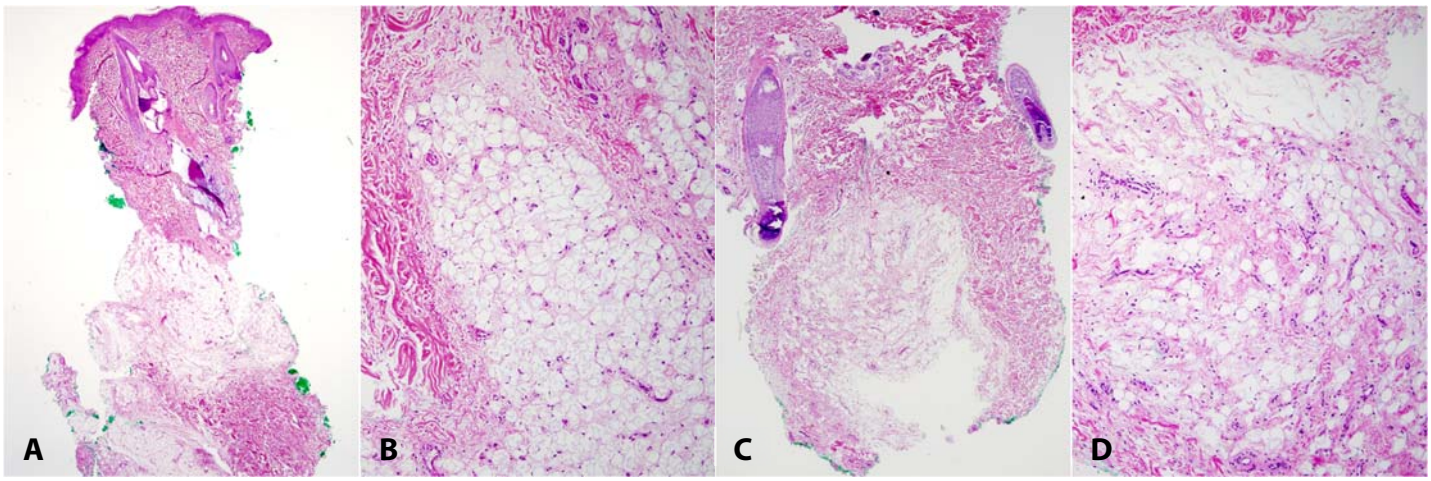

Figure 2. *Non-scarring alopecia with minimal lymphocytic inflammation and lipoatrophic changes of the subcutaneous tissue. H&E,* ***A)*** *40×, and* ***B)*** *100×. Non-scarring alopecia with reduced density of terminal hair follicles and small adipocytes associated with mildly increased vasculature and fibrous tissue. H&E,* ***C)*** *40× and* ***D)*** *100×.*

have failed to alleviate lipedematous alopecia include coal tar shampoo and hydroxychloroquine [5]. In one case of lipedematous alopecia in the setting of scalp psoriasis, topical steroids resulted in reversal of alopecia [2]. Success with mycophenolate mofetil, finasteride, and systemic corticosteroids has also been reported [1,5,7].

The differential diagnosis in lipedematous alopecia includes lipedematous scalp, lupus panniculitis, benign lipoma (specifically the spindle-cell variant), alopecia areata, and cutis verticis gyrata. Lipedematous scalp is similar to lipedematous alopecia in that there is expansion of the subcutaneous tissue. However, there is no associated hair abnormality [3]. Lupus erythematosus panniculitis of the scalp typically presents clinically with a linear geometry and is histologically characterized by fat necrosis, presence of abundant mucin in fat lobules, and lymphocytic infiltrate [8]. Spindle cell lipomas classically occur on the shoulder or neck of an adult male, but have been reported on the scalp [9]. Histologic findings in spindle cell lipoma include clusters of spindled-cells and mast cells lacking a lymphocytic infiltrate [9]. Alopecia areata (AA) is clinically characterized by discrete areas of smooth alopecia with fractured "exclamation point hairs" on gross examination. Further, the increased adiposity seen on microscopy in our patient would not be consistent with the diagnosis of AA; typically in AA an increased proportion of telogen and categen hairs is expected [10]. Cutis verticis gyrata (CVG) is characterized by cerebriform furrows and folds and can be classified as primary essential CVG with no underlying abnormalities or secondary CVG with associated systemic findings. A defining histopathological finding in CVG is sebaceous gland hypertrophy. In lipedematous alopecia there is no gross undulating pattern of the affected area and no sebaceous gland hypertrophy [3].

Conclusion

Our patient had clinical and histological features consistent with lipedematous alopecia, a boggy, erythematous plaque with alopecia of the occipital scalp and subcutaneous thickening with lymphocytic dermal infiltrate and decreased anagen hairs on histology. This case emphasizes the importance of careful clinical-pathological correlation in the diagnosis of lipedematous alopecia and also highlights the broad demographic at risk for lipedematous alopecia, spanning age, gender, and ethnicity.

Potential conflicts of interest

The authors declare no conflicts of interests.

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