CASE REPORT

Lipedematous alopecia with mucinosis: report of the first case in Taiwan

Jui-Hung Ko 1,1, Yi-Chin Shih 1,4,†, Cheng Hong Toh 2,4, Hua-En Lee 1,4, Tseng-tong Kuo 3,4, Rosaline Chung-Yee Hui 1,4,*

1 Department of Dermatology, Chang Gung Memorial Hospital, Taipei, Taiwan
2 Department of Diagnostic Radiology, Chang Gung Memorial Hospital, Taipei, Taiwan
3 Department of Pathology, Chang Gung Memorial Hospital, Taipei, Taiwan
4 Chang Gung University College of Medicine, Taiwan

A R T I C L E  I N F O

Article history:
Received: Jan 25, 2010
Revised: Mar 29, 2010
Accepted: Apr 27, 2010

Keywords:
Lipedematous alopecia
Lipedematous scalp
Mucinosis

A B S T R A C T

Lipedematous alopecia is an uncommon disease that mainly affects African American and Egyptian women.1,2 It is characterized by thickened subcutaneous tissue in the scalp with varying severity of hair loss.3 This report is of an 18-year-old Taiwanese woman who presented with asymptomatic boggy and thickened scalp for 10 years. In the last 6 months, there was diffuse hair loss on the affected scalp without scarring. Histopathologically, there were increased thickness of subcutaneous fat layer, mild perivascular lymphocytic infiltration, and separated collagen bundles in the dermis. Alcian blue stain demonstrated mucin deposition in the dermis and subcutis, whereas magnetic resonance imaging showed thickened scalp with expanded subcutaneous fat layer. The clinical findings and imaging study established the diagnosis of lipedematous alopecia. The pathogenesis and disease etiology remain unclear. The coexistence of mucin is extremely rare and its significance should be further investigated.

Copyright © 2011, Taiwanese Dermatological Association. Published by Elsevier Taiwan LLC. All rights reserved.

Introduction

Lipedematous alopecia is a rare disorder mainly affecting African American and Egyptian women.1,2 It is characterized by thickened subcutaneous tissue in the scalp with varying severity of hair loss.3 This report is of an 18-year-old young female with lipedematous alopecia and an unusual histopathologic finding of mucin deposit.1,4 Thus far, this is the first case of lipedematous alopecia with mucinosis reported in Taiwan.

Case report

An 18-year-old woman presented with asymptomatic boggy and diffuse swelling scalp with spongy consistency affecting the vertex, bilateral parietal, and occipital areas for more than 10 years. She denied trauma or medication history. In the last 6 months, there was a note of gradual diffuse hair loss with hair thinning in regions of the boggy scalp. There is no family history of similar condition and no change in diet or psychosocial stress.

On physical examination, the scalp was soft, thickened, and boggy without erythema, scarring, or pustules. Aside from the vertex, the occipital and parietal areas showed reduced hair density compared with the temporal areas (Figures 1A and 1B). Hair pull test revealed shedding of less than two hairs per hair pull on three instances.

Laboratory data, including complete blood cell count, thyroid function test (i.e. free thyroxine and thyroid-stimulating hormone), free testosterone, dihydroepiandrosterone sulfate, antinuclear antibody, rapid plasma reagin, and hemoglobin A1c were all within normal limits. Plasma protein electrophoresis showed no paraprotein. Biopsy was taken and histopathology showed an expansion of the subcutaneous fat layer. Most of the hair follicles were in the anagen phase, and there were separated collagen fibers with a few lymphocytes infiltrating the dermis (Figures 2A and 2B).

Increased intercollagen space hinted at the possibility of deposition disease. Alcian blue stain with hyaluronidase demonstrated mucin deposits in the dermis and subcutis (Figures 2C and 2D). Ultrasonography showed that the thickness of the vertex scalp (1.04 cm) was nearly twice that of normal individuals (Figure 3), whereas magnetic resonance imaging (MRI) also showed thickened scalp (16.93 mm) with expansion of the subcutaneous fat layer (Figure 4). In contrast, normal scalp thickness of the vertex was 6.4 ± 1.21 mm (mean ± SD, range, 4.37–8.34 mm; Figure 5) by MRI of 10 young Taiwanese females (mean age, 23.6 years; range, 15–29 years). This confirmed the diagnosis of lipedematous scalp, but the presence of mucin, ruptured hair follicles, and inflammation with hair loss suggested the final diagnosis of lipedematous alopecia.
Lipedematous scalp was first described by Cornbleet\(^5\) in 1935 in a 44-year-old black woman with cotton batting-like feeling on her scalp without any hair abnormality. Skin biopsy was unremarkable except for expansion of the subcutaneous fat layer. Lipedematous alopecia was termed by Coskey et al.\(^6\) in 1961 when they reported two cases of shortened hairs with length < 2 cm associated with increased thickness of the scalp subcutaneous fat layer. In 1994, Lee et al.\(^7\) coined the term lipedematous scalp in a 32-year-old black woman with a similar description as Cornbleet’s\(^5\) case. Lipedematous alopecia and lipedematous scalp both share characteristic thickened scalp and differ only in hair abnormalities.\(^8\)

Various methods are used to measure scalp thickness, including introducing sterile needles, ultrasonography, computed tomography, and MRI. The thickness of lipedematous scalp ranges from 9 to 22 mm, with an average of 13.2 mm.\(^1,3,4,6–24\) In comparison, the average scalp thickness of 523 healthy adults is 5.8 ± 0.12 mm using roentgenographic measurements.\(^25\)

To date, there are 21 cases of lipedematous alopecia and 20 cases of lipedematous scalp, including the current case, reported in the literature.\(^1–24\) Most cases are African Americans and Egyptians, with only three Asians.\(^7,13,16\) There is female preponderance (93%) with only four male patients reported.\(^13,18,23\) Lipedematous alopecia and lipedematous scalp are acquired, late-onset diseases with median age of onset of 48 years (range, 6 months–83 years) and median duration of disease before diagnosis is 2 years (range, 2 months–15 years). Scalp thickness ranges from 9 to 22 mm, with an average of 13.2 mm.\(^1,3,4,6–24\)

**Figure 1** (A) Physical examination showed diffuse hair loss with reduced hair density on the vertex, parietal, and occipital areas. (B) The involved scalp was boggy and thickened with spongy consistency, as shown by the pinched soft area.

**Figure 2** (A) There was increased thickness of the subcutaneous fat tissue layer, with many anagen hair follicles and a few perivascular lymphocytic infiltrate in the dermis (H&E, original magnification 200×). (B) There were separated collagen fibers in the dermis (H&E, original magnification 200×). (C) There were mucin deposits in the dermis, and (D) the subcutis (yellow arrows) (H&E, original magnification 200×). H&E = hematoxylin and eosin stain.
Clinically, lipedematous alopecia is characterized by boggy and thickened scalp with spongy or jelly-like consistency.\(^1,18,22\) Hair abnormalities include diffuse hair loss or inability to grow long hairs.\(^18\) Most patients are asymptomatic although some complain of painful sensation, paresthesia, mild itch, and cotton wool-like feeling over the scalp.\(^3,6,9,10,17,23\) Most affected regions are on the vertex and occipital areas, with few cases centering on the parietal and frontal areas.\(^14\) and may even progress to the entire scalp in some cases.\(^7,17\)

Histopathologically, thickened subcutaneous adipose tissue composed of mature fat cells without cellular or nuclear anomalies is the consistent feature.\(^12,20,23\) The surrounding hair follicles and adnexal structures are intact. Some cases have perivascular lymphocytic infiltrates in the dermis.\(^9,10,23\) Other inconsistent histopathologic findings included hyperkeratosis,\(^5,6,15,23\) focal spongiosis,\(^3\) elastic fibers fragmentation,\(^14\) collagen bundle degeneration,\(^23\) dilated lymphatic vessels in the dermis,\(^3,21\) edematous change in the dermis and subcutis,\(^2,3,12\) and perifollicular fibrosis.\(^5,10\) Most cases do not show mucin deposition in the dermis or subcutis, with only one case showing glycosaminoglycan deposition by Alcian blue stain at pH 2.5.\(^10\) The current case demonstrates mucin in the dermis and subcutis, and the relationship between lipedematous alopecia and mucinosis warrants further investigation.

The etiology and pathogenesis remain unclear. Fair et al\(^12\) suggest that localized edema with degeneration and disruption of adipose tissue may be a possible cause. Martin et al\(^3\) and Morawej-Farshi and Mohtasham\(^21\) report three cases of lipedematous alopecia with characteristics of lymphatic dilatation in the dermis. This phenomenon may explain the consequences of edematous change.

Thickening of the subcutaneous fat has been considered as a secondary response. However, there is no evidence of lymphatic dilatation in pathology or history of using headwear in the current case. Although original cases are largely reported in African American women, more recent reports of Egyptians, Asians, and European Caucasians\(^14,18\) suggest a more heterogeneous racial distribution. Hormonal factor is also suspected as an important cause, because lipedematous alopecia affects predominantly female patients. However, Ikejima et al\(^13\) propose that the disease may be underreported in male patients, suggesting that some cases may be misdiagnosed as androgenetic alopecia.

Lipedematous alopecia and lipedematous scalp should be differentiated from cutis verticis gyrata, which is characterized by asymptomatic thickening of the scalp where sparse hairs grow over the fold and normal hair density is in the furrows (folds and furrows pattern).\(^8\) MRI shows increased thickness in the dermis and slight expansion of the subcutaneous fat layer.

There is no satisfactory treatment. Coskey et al\(^6\) report partial response with systemic steroids. Intraleional steroids have been...
tried in Curtis and Heising's case but without improvement.9
Topical steroids also provide no improvement.11,17 In the current
case, topical and intrallesional steroids show no effect on boggy
scalp. There is one reported case of disease progression after
surgical excision of the boggy scalp.15

In summary, there are only three reported cases of lipedematous
alopecia and lipedematous scalp involving Asians.7,13,16 However,
the diagnosis is not easily recognized and may therefore be
underreported. Diagnosis of lipedematous alopecia is dependent on
clinical manifestations and imaging studies. Biopsy specimens are
usually not deep enough to represent the whole subcutaneous
adipose layer, but can exclude differential diagnoses. The presence
of mucin deposit, as in the current case, has been previously
reported only once. Hence, its association and signiﬁcance need
further investigation. Increased awareness of lipedematous scalp
and alopecia should lead to more reporting and better delineation.

References

uncommon clinicopathologic variant of nonscarring but permanent alopecia.
2. El Darouti MA, Marzouk SA, Mashaly HM, et al. Lipedema and lipedematous
3. Martin JM, Monteagudo C, Montesinos E, et al. Lipedematous scalp and lipe-
dematous alopecia: a clinical and histologic analysis of 3 cases. J Am Acad
Actas Dermosifiliogr 2009;100:69–72. [In Spanish]
688.
619–22.
7. Lee JH, Sung YH, Yoon JS, Park JK. Lipedematous scalp. Arch Dermatol 1994;130:
802–3.
9. Curtis JW, Heising RA. Lipedematous alopecia associated with skin hyper-
15. Scheuﬂer O, Kania NM, Heinrichs CM, Exner K. Hyperplasia of the subcutaneous
adipose tissue is the primary histopathologic abnormality in lipedematous scalp.
17. High WA, Hoang MP. Lipedematous alopecia: an unusual sequela of discoid
20. Mansur AT, Yasar S, Aydingoz IE, et al. Colocalization of lipedematous scalp and
25. Carro SM, Selby S, Young R. Scalp thickness and the fat-loss theory of balding,
AMA Arch Derm Syphilol 1954;70:601–8.