Lipedematous Scalp and Alopecia: Report of Two Cases with a Brief Review of Literature

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Abstract

Lipedematous scalp (LS) and lipedematous alopecia (LA) are both rare conditions with an unknown etiology. LS is characterized by boggy swelling under the skin as a result of hyperplasia of subcutaneous layer. LA is basically LS associated with hair growth abnormalities such as alopecia and short broken hair. Herein, we present two patients who were diagnosed with LS and LA where case with LA had a new diagnosis of systemic lupus erythematosus.

Key Words: Autoimmunity, lipedematous alopecia, lipedematous scalp, lupus erythematosus

Introduction

Lipedematous scalp (LS) is a rare disease characterized by boggy swelling of the skin as a result of increased subcutaneous layer thickness. The term lipedematous alopecia (LA) is used when hair growth abnormalities such as alopecia and short broken hair are coexistent with LS. LS is often asymptomatic and incidentally detected whereas LA is frequently reported to cause pruritus, pain, and paresthesia. Hyperelasticity of skin, laxity of joints, renal failure, diabetes mellitus, and discoid lupus erythematosus (DLE) have been described as associated medical conditions. Herein, we present two patients who were diagnosed with LS and LA where the case with LA was also suffering from systemic lupus erythematosus (SLE), hitherto unreported.

Case Reports

Case 1

A 33-year-old female patient attended the dermatology outpatient clinic with a complaint of diffuse, cotton-like swelling of scalp which was present for a year. She described that the swelling started from vertex and spread throughout the whole scalp. Medical and family history was unremarkable. On dermatological examination, the scalp was thickened and palpation of the skin revealed soft, boggy, and spongy texture without fluctuation [Figure 1a]. On high-frequency skin ultrasound imaging (HF-USG), compressible massive soft-tissue edema was noted. Magnetic resonance imaging showed diffuse thickening of subcutaneous fat tissue with a thickness of 12 mm. A biopsy was taken from the scalp, and histopathological examination reported increased thickness of normal subcutaneous adipose tissue consistent with LS [Figure 1b]. Since the patient was not disturbed by her skin condition, she was advised regular follow-up without any medical intervention at that time.

Case 2

A 36-year-old female patient attended to the dermatology outpatient clinic with complaints of thickening of scalp and associated hair loss for about 2 months. The thickening was first noted on vertex and spread to posterolateral aspects of the scalp. She also complained of hyperesthesia and alopecia over the affected part of the scalp. On medical history, a prior diagnosis of undifferentiated connective tissue disease (CTD) was present, but the patient had refused...
to take treatment and had not applied for follow-up for this condition. On dermatological examination, soft, boggy swelling on the scalp was palpated, especially on vertex, parietal, and occipital areas. Alopecic plaques with 2cm×1cm to 4cm×1cm dimensions were detected where follicular ostia were easily noted without erythema, scaling, or scarring. Brittle white terminal hair was also observed on peripheral sites of these patches [Figure 2a]. In addition, the patient had mild erythema with fine telangiectasias on bilateral malar area and described photosensitivity which was present for last few years. On laboratory workup, complete blood count, full biochemistry, and hormone levels (TSH, fT3, fT4) were within normal ranges whereas serum vitamin B12 level was low, i.e., 99 pg/mL (126–590 pg/mL) and antinuclear antibody (ANA) was positive at a titer of 1 in 100. HF-USG showed marked thickening of subcutaneous tissue particularly on plaques with hair loss where the distinction of epidermis, dermis, and adipose tissue disappeared [Figure 2b]. Computed tomography showed increase in subcutaneous fat tissue; the thickness was measured 11 mm on vertex. A biopsy was taken from the scalp revealing ectatic vascular structures together with mild perivascular inflammation composed of lymphocytes and eosinophils [Figure 2c]. The patient consulted the rheumatology department for malar erythema, ANA positivity, and previous diagnosis of undifferentiated CTD. On rheumatologic evaluation, anti-SS-A (anti-Ro) positivity was detected on extractable nuclear antigen panel. She was diagnosed with SLE, and a treatment regimen of hydroxychloroquine 2 × 200 mg/day and azathioprine 3 × 50 mg/day was recommended. However, the patient again refused to receive any systemic treatment. Nevertheless, spontaneous hair growth was reported by the patient despite lack of treatment.

Discussion

LS and LA are two rare entities which are characterized by thick, boggy, cotton-like swelling of the scalp. Based on related literature, there are a total of 43 LS and 38 LA patients reported [Table 1]. LA/LS can begin at any age, and in fact congenital cases have been reported.[1] There is a remarkable female preponderance (F:M=67:14) for the disease, and although pathogenesis of LS/LA is not well defined, hormonal factors are believed to play a role because of increased frequency in female gender. Leptin disregulation is claimed to play a role in the pathogenesis of LA/LS.[2,3] Detailed genetic factors have not been explained for LS/LA, and to date, only two LS cases were reported with family history involving mother and daughter.[4,5] Hair loss observed in LA is thought to be induced by pressure on hair follicles caused by thickening of subcutaneous tissue. This pressure may lead to decreased hair growth or shortening of anagen period of the hair follicle.[5] Lymphatic vessel dilatation which was also seen
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Table 1: Characteristics of patients diagnosed with lipedematous scalp and lipedematous alopecia

| Author/year   | Diagnosis | Cases; sex/age | Coexisting diseases | Histopathological findings |
|---------------|-----------|----------------|--------------------|---------------------------|
| Cornbleet/1935 | LS        | Female/44      | -                  | Moderate hyperkeratosis and follicular plugging, atrophic follicles replaced by fibrous tissue, free hair fragments of hair follicle on dermis, increased fat tissue |
| Coskey/1961   | LA        | Female/29, Female/75 | Diabetes mellitus | Hyperkeratosis, patchy mid-dermal lymphocytic infiltrate, increased subcutaneous fat |
| Curtis/1984   | LA        | Female/62      | Skin and joint hyperelasticity | Minimal inflammation with mild perifollicular fibrosis, increased mucin |
| Lee/1994      | LA        | Female/32      | -                  | Mild hyperkeratosis and acanthosis, follicular plugging |
| Kane/1998     | LA        | Female/49      | -                  | Mild basket-weave hyperkeratosis, superficial perivascular lymphocytic infiltrate, increased thickness of the subcutaneous fat layer |
| Fair/2000     | LS        | Female/18      | -                  | Increased thickness of subcutaneous adipose tissue, marked decrease in the number of hair follicles |
| Bridges/2000  | LA        | Female/48      | Renal insufficiency | Focal edema, fragmentation of elastic fibers |
| Curtis/1984   | LA        | Female/62      | Skin and joint hyperelasticity | Hyperkeratosis, increased subcutaneous fat without cellular or nuclear anomalies |
| Lee/1994      | LA        | Female/32      | -                  | Mild hyperkeratosis and acanthosis, follicular plugging |
| Kane/1998     | LA        | Female/49      | -                  | Mild basket-weave hyperkeratosis, superficial perivascular lymphocytic infiltrate, increased thickness of the subcutaneous fat layer |
| High/2005     | LA, LS    | Female/57, female/55 | Discoid lupus erythematosus | Parakeratosis, follicular plugging, superficial and deep perivascular/periadnexal lymphocytic infiltration, expansion of subcutaneous tissue without alopecia |
| Mansur/2006   | LS        | Female/46      | Nevis lipedematous superficialis | Thinning of dermis and increase in the subcutaneous fat tissue are seen on the soft area peripheral to nevus lipomatosus superficialis; bulbi of anagen hair are embedded in the fatty tissue |
| Piracini/2006 | LA        | Male/48, male/53 | Androgenetic alopecia | Thickened adipose tissue with reduced terminal to vellus hair ratio |
| Rowan/2006    | LS        | Female/9       | -                  | Thickenened mature subcutaneous adipose tissue with disruption of fatty architecture |
| Yasar/2007    | LA, LS    | Female/45, male/49, female/62 | Positive family story | Epidermal atrophy, coarse collagen bundles with collagen degeneration in superficial dermis, mild perivascular lymphocytic infiltration, hyperkeratosis, epidermal atrophy, a scant perivascular mononuclear infiltrate |
| El Darouti/2007 | LA/LS | 10 cases        | -                  | Thicknessed subcutaneous tissue with normal morphology of the hair follicles and epidermis |
| Martinez/2007 | LS        | Female/77      | -                  | Increased thickness of the subcutaneous adipose tissue with dilated lymphatic vessels in dermis |
| Farshi/2007   | LA        | Female/45      | Cafe au lait spots | Enlargement of subcutaneous fat layer, dilated lymphatic vessels in upper dermis |
| Torres/2008   | LA        | Female/55      | -                  | Loss of septa between fatty lobes, vertical fibrosis structures replacing preexisting hair follicles |
| Gonzalez/2008 | LA        | Female/52      | -                  | Dispersed fat cells in the middle to deep reticular dermis with loss of hair follicles; central fibrous tract in the deep reticular dermis showing vertically orientated fibrous tissue surrounded by adjacent fat cells |
| Yip/2008      | LA        | Female/67      | -                  | Mild perivascular lymphocytic infiltration and marked hyperplasia of subcutaneous tissue |

Contd...
in our patient is also suggested to play a role in alopecia in some cases.\cite{5}

Scalp thickening is usually reported to start from vertex and occipital areas and slowly expand to the entire scalp which was also described by our patients. Local symptoms such as pruritus, paresthesia, and headache have been reported on affected areas and are more prevalent in LA.\cite{1,2}

There are LA/LS cases reported with medical conditions such as skin and joint hyperelasticity, kidney failure, scalp psoriasis, and breast cancer.\cite{6,7} However, these conditions seem to be coincidental; hence, there is no demonstrated relationship between LA/LS and these disorders. Cases with coexistent autoimmune connective tissue disorders such as Sjogren’s syndrome and DLE raise suspicion for a common pathogenetic pathway due to the resemblance of histopathological features of LA with LE.\cite{8} Although increased subcutaneous fat layer is observed in all cases of LS/LA, additional findings are reported in histopathological specimens (Table 1). These include dilated lymphatic vessels, dermal edema, perivascular lymphocytic infiltration, and mild perifollicular fibrosis. For instance, perivascular, periadnexal lymphocytic infiltrate along with hyperkeratosis, follicular plugging, and fibrosis which can be seen in LA are common features that can be observed in late phases of DLE.\cite{8} Our LA patient who was later diagnosed with SLE supports this hypothesis; we think that the patient’s prior refusal to take a treatment for a diagnosis of CTD might have led to uncontrolled systemic inflammation and consequently to LA.

Different treatment modalities were used in LA/LS including intralesional and systemic steroids, surgical debulking, and other immunosuppressive agents such as mycophenolate mofetil. Treatment with debulking and mycophenolate mofetil was found successful in reported cases.\cite{2,9} Spontaneous hair growth may be observed despite lack of treatments as seen in our LA case.

In conclusion, LA/LS are both rare entities and share common histopathological features such as subcutaneous fat tissue enlargement, ectatic lymphatic vessels, and perivascular lymphocytic infiltrate which are seen in some autoimmune systemic diseases such as LE. Therefore, careful evaluation of LA/LS patients regarding coexistent inflammatory conditions may be helpful.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

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**Table 1: Contd...**

| Author/year       | Diagnosis | Cases; sex/age | Coexisting diseases | Histopathological findings                              |
|-------------------|-----------|----------------|---------------------|--------------------------------------------------------|
| Cunha Filho/2010  | LA        | Female/13      |                     | Focal inflammatory perifollicular infiltrate and discrete edema with ectatic lymphatic vessels |
| Yasar/2010        | LA/LS     | 31 cases       |                     | Dermal edema, elastic and collagen fiber fragmentation |
| Zeng/2011         | LS        | ND             | Heterochromia        | Thyickened subcutaneous fat layer with edema and lymphatic enlargement in the dermis |
| Ko/2011           | LA        | Female/18      |                     | Hyperplasia of the subcutaneous fat without cellular or nuclear anomalies |
| Muller/2015       | LA        | Male/15        |                     | Mild perivascular lymphocytic infiltration, dermal edema, enlargement of subcutaneous fat tissue |
| Fuentelsaz/2012   | LA        | Female/49      | Psoriasis, breast cancer | Thickenened mature subcutaneous fat with a decreased number of hair follicles |
| Lee/2015          | LA        | Female/6, female/10 |                     | Superficial perivascular lymphocytic infiltrate with dilated lymphatic vessels, mature adipocytes with loss of normal architecture due to interstitial edema |
| Cabrera/2015      | LA        | Male/51        | -                   | Thickenened subcutaneous tissue without inflammatory infiltrate |
| Carasco-Zuber/2016| LS        | Female/48      | Hypertension         | Thickened subcutaneous tissue, perivascular lymphocytic infiltration, ectatic lymphatic vessels |
| Wang/2016         | LA        | Male/20        |                     | Mild perivascular mononuclear infiltration and perifollicular fibrosis in dermis with mucin deposition |
| Present cases     | LS, LA    | Female/33, female/36 | SLE | Thickenened subcutaneous tissue, perivascular lymphocytic infiltration, ectatic lymphatic vessels |

NP: Not performed, LA: Lipedematous alopecia, LS: Lipedematous scalp, SLE: Systemic lupus erythematosus, ND: Not determined


**Conflicts of interest**

There are no conflicts of interest.

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