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CASE REPORT

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

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ABSTRACT

Lipedematous alopecia is an uncommon disease that mainly affects African American and Egyptian women. 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.

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 compete 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 A_{1c} 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 $\pm$ 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.

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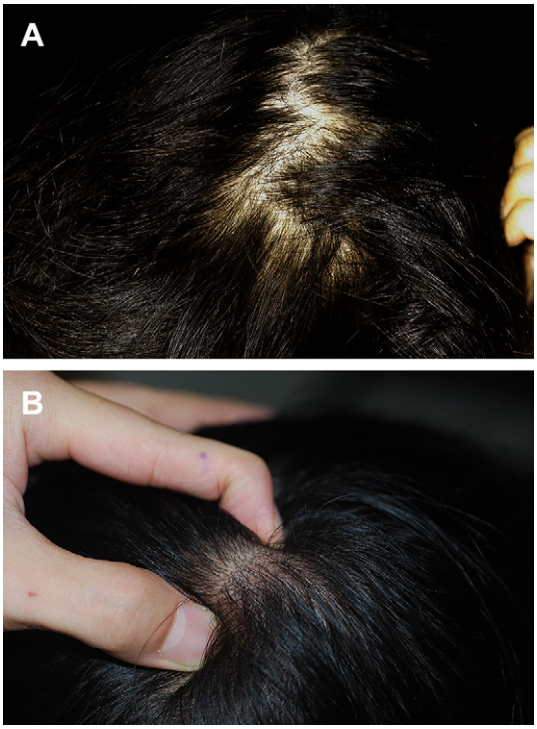

Figure 1 (A) Physical examination showed diffuse hair loss with reduced hair density on the vertex, parietal, and occipital areas. (B) The involved scalp was boggy and thickened with spongy consistency, as shown by the pinched soft area.

Discussion

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]

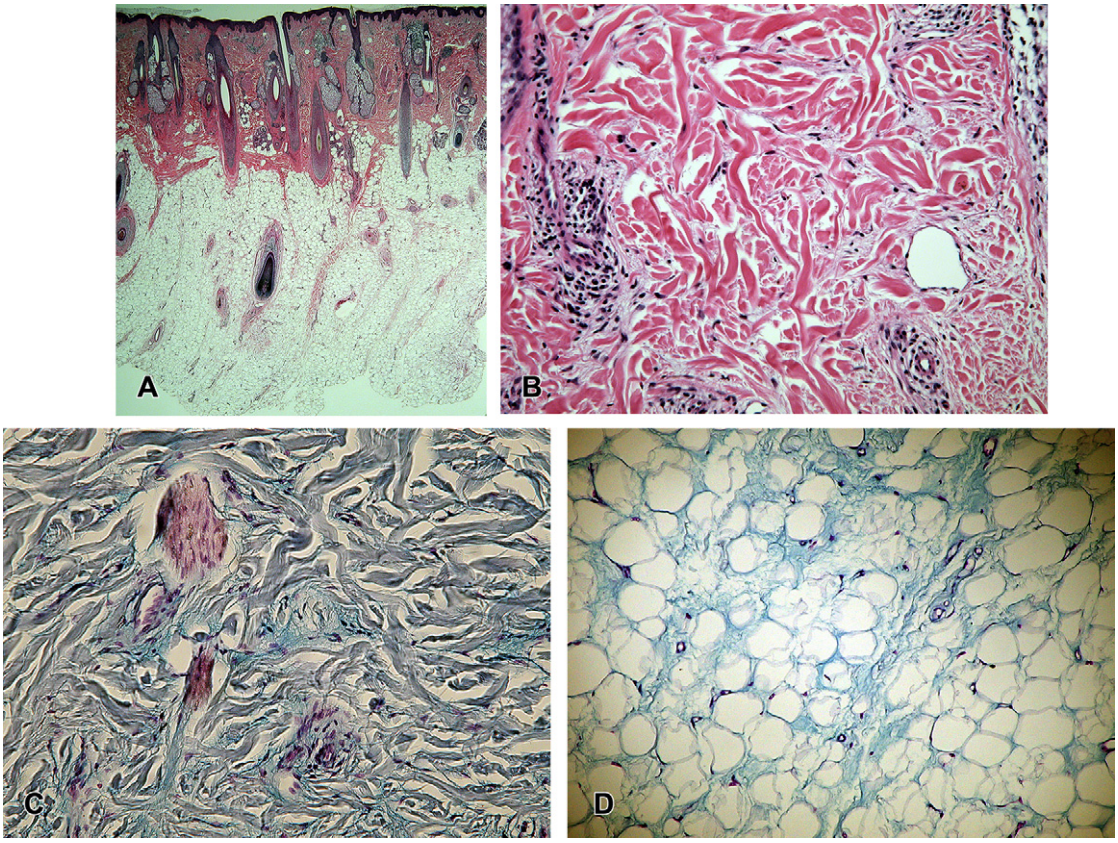

Figure 2 (A) There was increased thickness of the subcutaneous fat tissue layer, with many anagen hair follicles and a few perivascular lymphocytic infiltrate in the dermis (H&E, original magnification 20×). (B) There were separated collagen fibers in the dermis (H&E, original magnification 200×). (C) There were mucin deposits in the dermis, and (D) the subcutis (yellow arrows) (H&E, original magnification 200×). H&E = hematoxylin and eosin stain.