Lipedematous scalp in a patient with ankylosing spondylitis

**Maria Gabriela Lang, Mauro Waldemar Keiserian,
Rodrigo Pereira Duquia**

ABSTRACT

The lipedematous scalp is a rare condition of unknown etiology characterized by diffuse increase of the fat layer in the subcutaneous tissue of the scalp. Case Report: We described the case of a patient with ankylosing spondylitis treated with etanercept and prednisone, who develops diffuse thickening of the scalp associated with local pain, without improvement with analgesics and non-steroidal anti-inflammatory drugs. The previous cases reported described no clear association between comorbidities or medication use and development of this disease. Conclusion: There is no effective treatment for this condition, but is reported the successful use of mycophenolate mofetil for lipedematous alopecia, variant of lipedematous scalp.

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CASE REPORT

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Introduction: The lipedematous scalp is a rare condition of unknown etiology characterized by diffuse increase of the fat layer in the subcutaneous tissue of the scalp. **Case Report:** We described the case of a patient with ankylosing spondylitis treated with etanercept and prednisone, who develops diffuse thickening of the scalp associated with local pain, without improvement with analgesics and non-steroidal anti-inflammatory drugs. The previous cases reported described no clear association between comorbidities or medication use and development of this disease. **Conclusion:** There is no effective treatment for this condition, but is reported the successful use of mycophenolate mofetil for lipedematous alopecia, variant of lipedematous scalp.

Keywords: Ankylosing spondylitis, Autoimmune disease, Lipedematous scalp, Scalp dermatoses

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INTRODUCTION

The lipedematous scalp was first described by Cornbleet in 1935 [1]. This entity of unknown etiology is described as thickening of the scalp subcutaneous tissue without capillary changes, usually accompanied by local pain and less commonly by paresthesia and pruritus [2]. The lipedematous alopecia is a similar disease associated with varying degrees of hair loss and/or inability to maintain hair growth above 2 cm, although there is no consensus if they can be spectra of the same disease [3, 4].

The objective of this study is to report the case of a patient with ankylosing spondylitis who showed clinical and histological features compatible with lipedematous scalp.

CASE REPORT

A 63-year-old white female started about three months earlier to feel pain like grip in the scalp worsened by touching. She noticed diffuse scalp thickening. The patient was previously diagnosed with ankylosing spondylitis and

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was positive for HLA B27. She was in use of etanercept 50 mg weekly since 2011 and prednisone 7.5 mg/day, with no joint or axial complaints. The patient denied hair pulling or excessive manipulation of the hair or scalp, local trauma, use of topical treatment or family history of similar condition. She did not notice any hair changes. Physical examination revealed diffuse thickening of the scalp with consistency of layered gauze or cotton batting, painful on palpation. There was no visual evidence of local inflammatory signs or increased hair fragility. The “pull test” was within normal limits and no abnormalities of the hair shaft were seen by light microscopic examination. Complete blood cell count and biochemical parameters were within normal limits. Metabolic screening was negative, inflammatory tests were normal, serum protein electrophoresis showed no paraprotein and research of antinuclear antibodies (ANA) and rheumatoid factor were negative. Magnetic resonance imaging (MRI) scan with focus to the scalp showed diffuse thickening of subcutaneous tissue involving the scalp, measuring 17 mm in the occipital region. The study of brain parenchyma showed a small area of hyperintensity on T2 without restriction on broadcasting in fluid attenuated inversion recovery (FLAIR) sequence in periventricular white matter consistent with microangiopathy (Figure 1). Histopathological analysis of the scalp showed thickening of the subcutaneous tissue with relative thinning of the dermis, marked edema of the dermis, vascular ectasia with discrete lymphocytic infiltrate (Figure 2). She was treated with common analgesics and NSAIDs, but no improvement of symptoms was noticed and there was no change in the thickness of the scalp, which remained stable.

DISCUSSION

We described a patient with ankylosing spondylitis who developed lipedematous scalp. There is no clear association between lipedematous scalp and a specific medical disorder and its pathogenesis remains unclear [2, 5]. There is ongoing debate involving hormonal factors, since most of the reported cases occurred in women and is proposed an association with obesity [3, 5, 6]. There is no clear association between this pathology and medical or physiologic conditions, but lipedematous alopecia was reported in a patient with skin and nail psoriasis, an autoimmune disease as the presented case [7]. The thickness of the normal scalp ranges from 0.4 to 6 mm [2]. This measurement can be performed by ultrasound, MRI, tomography or fine needle and the extent found in studies have ranged from 10–22 mm, compatible with the finding of 17 mm in the present case [3]. Histopathologic findings are nonspecific, being described thinning of the dermis, increased subcutaneous fat tissue extending to the superficial dermis in some cases, inflammatory infiltration with lymphocyte predominance, as demonstrated in

Figure 1: Magnetic resonance imaging (A) Sagittal scan of T2, (B) Axial scan of T2, (C) Axial scans of T1.

Figure 2: Thickening of subcutaneous tissue, dermis with relative thinning and marked edema, vascular ectasia with mild lymphocytic infiltrate (H&E stain, x100).

this study, ectatic lymph vessels [5, 6, 8]. Some showed hyperkeratosis, but the distribution of hair follicles is normal and that differentiates lipedematous scalp from lipedematous alopecia [3]. Few studies have described about the treatment and there is no consensus regarding treatment options. Most of the studies describe the use of non-steroidal anti-inflammatory drugs (NSAIDs) and symptomatic for complaints of numbness, pruritus and pain [7]. It was reported the use of hydroxychloroquine and topical corticosteroids with little improvement of symptoms and relapse in patients with lipedematous alopecia [7, 8].

Some patients were followed without any treatment, showing that some areas may become more thickened over the time [5]. Attempts with corticosteroids local infiltration did not showed influence in reducing scalp thickening [9]. Recently, a case report described the use of mycophenolate mofetil with thickness reduction of scalp and improvement hair growth and density in lipedematous alopecia [10]. Additional studies about the pathophysiology, risk factors and mechanisms involved in its appearance are needed, so that specific therapeutic approach could be developed because of the rarity of this entity.

CONCLUSION

The lipedematous scalp is still a disease that remains with its pathophysiology poorly defined. This patient had an autoimmune disease and it might serve as a trigger for the disease because it was already demonstrated an association between lipedematous alopecia, a very similar condition, and psoriasis also, in a few cases was noted some improvement with the use of mycophenolate mofetil.

Author Contributions

Maria Gabriela Lang – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Mauro Waldemar Keiserman – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Rodrigo Pereira Duquia – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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