Skin Appendage Disord 2017;3:92–94
DOI: 10.1159/000462980**Atypical Localization of Necrobiosis Lipoidica:
Involvement of the Face and Scalp**Şirin Yaşar^a Emre Kaynak^a Pembegül Güneş^b Fatih Göktay^a
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To the Editor,

Necrobiosis lipoidica (NL) is a chronic granulomatous dermatitis typically located on the lower extremities [1]. It was previously known as necrobiosis lipoidica diabetorum; however, as only a small number of patients with the condition have diabetes (0.3–1.2%), the term NL is now accepted. Forman [2] first reported NL of the scalp in 1954. Later, Jones et al. [3] reported atypical NL (ANL) of the face and scalp in 29 patients in the largest ANL study. However, NL does not come to mind in the first differential diagnosis of scarring alopecia.

A 44-year-old woman presented with an 8-month history of two atrophic, alopecic, and a few sclerotic lesions on the face and scalp. In the dermatological examination, two round plaques with raised, sclerotic, and telangiectatic alopecic plaques that were 2 × 3 and 1 × 1 cm in size were observed. Alopecic plaques were also seen on the frontal hairline and on the scalp (Fig. 1a–c). Laboratory tests obtained the following results: fasting blood sugar of 112–123–124 mg/dL (70–110), HbA1c value of 6% (4–6), the glucose tolerance test result in the first hour was 261 mg/dL (<200), and in the second hour it was 191 mg/dL (>140). The patient was referred to the diabetes polyclinic where she was diagnosed with type 2 diabetes mellitus for which she started treatment. A punch biopsy was taken from the sclerotic and alopecic areas. A histo-

pathological examination of the biopsy showed palisading granulomatous inflammation in the reticular and deep dermis with large numbers of multinucleate giant cells between collagen bundles; there was no mucin deposition seen with the periodic acid-Schiff (PAS) stain. Diminished hair follicles, perifollicular fibrosis, and perifollicular lymphocyte infiltration were detected (Fig. 2a–d). These findings were consistent with the diagnosis of NL. After the intralesional triamcinolone acetonide (5 mg/ml) was applied, the lesions began to diminish in size. Subsequently, systemic hydroxychloroquine (400 mg/day) and topical tacrolimus ointment (0.1% twice daily) were administered. This treatment resulted in moderate improvement at the end of the 1-year follow-up period and the lesions were softer and smaller (Fig. 3a–c).

NL is most commonly located in the pretibial area; it is rare on the trunk, upper extremities, and face/scalp. When these areas are involved, NL may be misdiagnosed as morphea, mycosis fungoides, granuloma annulare (GA), necrobiotic xanthogranuloma, actinic granuloma, discoid lupus erythematosus, or sarcoidosis.

Histopathologically, GA, cutaneous lupus erythematosus, sarcoidosis, actinic granuloma, and necrobiotic xanthogranuloma are considered in the differential diagnosis of ANL [4]. The diagnosis of GA is based on the presence of mucin deposition and the absence of necrobiosis. Patients with cutaneous lupus erythematosus have antinuclear antibodies positivity, and histological studies reveal basal vacuolar degeneration and interface changes. In addition, cutaneous sarcoidosis is a great mimicker and can present as atrophic and scarring alopecia [5]. Sarcoidosis has well-formed granulomas, lung involvement, higher serum angiotensin converting enzyme levels, and hypercalcemia and/or bone changes. Thus, these features may result in correct diagnoses and differential diagnoses of ANL. Solar degeneration and solar elastosis determinates, which are present in actinic granuloma, are absent in NL. In necrobiotic xanthogranuloma, histological tests reveal cholesterol clefts in the lesion (Table 1).

Fig. 1. a–c Annular plaques with raised, erythematous violaceous borders, sclerotic, telangiectatic, and alopecic plaque were seen on the anterior hairline and scalp.

Fig. 2. a–d Palisading granulomatous inflammation in the reticular and deep dermis with large numbers of multinucleate giant cells interdigitating between collagen bundles. No mucin was observed with PAS-Alcian blue stain (HE. $\times 40$, 100).

Fig. 3. a–c Softening and newly growing hair in the sclerotic plaque.

Table 1. Clinical, histopathological, and dermatoscopic features

	Clinical features	Histopathological features	Dermatoscopic features
NL	<ul style="list-style-type: none"> annular, nonscaling plaque mostly located on shins rarely, face and scalp involvement is seen 	<ul style="list-style-type: none"> interstitial and palisaded granulomas thickening of the blood vessel walls asteroid bodies are often found within giant cells. Loss of elastic tissue well-defined palisading granulomas are not present (contrary to sarcoidosis) absence of mucin deposition (present in GA) 	<ul style="list-style-type: none"> linear arborizing vessels a yellow background color, either alone or in combination with white or red yellow crusting ulceration [7]
GA	<ul style="list-style-type: none"> annular arrangement over distal extremities 	<ul style="list-style-type: none"> increased connective tissue mucins in GA 	<ul style="list-style-type: none"> vessel morphology is variable due to the different stages of disease progression [7]
Cutaneous lupus erythematosus	<ul style="list-style-type: none"> erythematous scaly plaques with follicular prominence scalp discoid lupus erythematosus presents as red, scaly, and bald plaques 	<ul style="list-style-type: none"> vacuolar interface dermatitis basement membrane thickening atrophy perifollicular fibrosis lymphoid infiltrate mucin deposition 	<ul style="list-style-type: none"> perifollicular whitish halo follicular keratotic plugs telangiectasias [8]
Morphea	<ul style="list-style-type: none"> depressed sclerotic plaque with an ivory white-colored center and erythematous border (“lilac ring”) 	<ul style="list-style-type: none"> abundant collagen deposits causing skin and appendage atrophy 	<ul style="list-style-type: none"> fibrotic beams crossed by spreading telangiectasias in the center of the lesion marked erythematous border [9]
Sarcoidosis	<ul style="list-style-type: none"> papules, nodules, plaques, erythema nodosum rarely scarring alopecia 	<ul style="list-style-type: none"> well-defined palisading granulomas “sarcoidal granulomas” 	<ul style="list-style-type: none"> “orange spots”, prominent telangiectasias dystrophic hairs [10]

NL, necrobiosis lipoidica; GA, granuloma annulare.

NL does not have an effective treatment, but topical or intralesional steroids may reduce the inflammation and sclerosis. Intralesional steroids, systemic hydroxychloroquine, and topical tacrolimus were used to treat the patient, resulting in moderate improvement. Although topical and intralesional steroids may reduce the acute inflammation phase, atrophy can increase in atrophic lesions in NL. Oral hydroxychloroquine has an anti-inflammatory action, it inhibits the prostaglandin synthesis, and shows an immunosuppressive action on macrophage migration, which can reduce inflammation in NL. Durupt et al. [6] reported that treatment with hydroxychloroquine and chloroquine had a good response in ANL within 3–6 months.

ANL should be considered in the case of alopecic and sclerotic lesions on the scalp and face. NL may easily be misdiagnosed as GA, actinic granuloma, lupus erythematosus, or cutaneous sarcoidosis. Although NL is rarely located on the scalp and is not considered in the prediagnosis of scalp lesions, detailed histopathological findings may help correctly diagnose NL.

Statement of Ethics

The patient's consent has been obtained.

Disclosure Statement

The authors have no conflicts of interest to disclose. There were no funding sources.

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