Necrobiosis Lipoidica on Skin Scars: A Case Report

Felipe Aguinaga1, Paula Regazzi de Gusmão1*, Alexandre de Almeida Filippo1 and Gustavo Filippo2

1Instituto de Dermatologia Prof. Rubem David Azulay, RJ, Brazil
2Universidade Estácio de Sá, RJ, Brazil

Abstract
Necrobiosis Lipoidica (NL) is an idiopathic granulomatous disease typically associated with diabetes. Classic lesions present as yellowish erythematous atrophic plaques, with telangiectasias, most frequently located in the pretibial region. We present a case of a 47-year-old, non-diabetic female patient with NL lesions appearing over surgical scars, after silicone breast implant surgery, and also over a scar from a childhood bicycle accident. NL occurring on pre-existing surgical scars has been reported in the literature. Among the possible pathogenic mechanisms, trauma seems to serve as a trigger for the development of NL. The appearance of lesions in pre-existing surgical scars has been interpreted by most authors as an example of Koebner’s isomorphic phenomenon. Our case highlights a rare presentation of NL, and reinforces the theory that trauma plays a role in the pathogenesis of this disease.

Introduction
Necrobiosis Lipoidica (NL) is an idiopathic granulomatous disease typically associated with diabetes [1]. Classic lesions present as yellowish erythematous atrophic plaques, with telangiectasias, most frequently located in the pretibial region. The appearance of NL lesions over surgical scars has been reported in the literature, and is generally understood as a form of Koebner’s isomorphic phenomenon, that is, the appearance of new skin lesions of a pre-existing dermatosis on areas of cutaneous injury in otherwise healthy skin [2]. We present a case of a 47-year-old, non-diabetic female patient with NL lesions appearing over surgical scars, 13 years after silicone breast implant surgery, and also over a scar from a childhood bicycle accident.

Case Report
A 47-year-old female patient presented two yellowish erythematous plaques with well-delimited borders, atrophic center and telangiectasias in the left breast for 2 years (Figure 1). The largest lesion, located in the infra-areolar scar measured about 10 cm in the largest diameter, and the smallest lesion in the superomedial portion of the periareolar scar measured about 3 cm in the largest diameter. During physical examination, yellowish erythematous papular lesions were observed on the right forearm, which had not been previously noticed by the patient (Figure 2). All lesions were adjacent to scars and, according to the patient, presented slow growth and no local symptoms. She reported that the scars on the breasts were from a silicone breast implant surgery performed 15 years earlier, and the scar on the forearm resulted from a trauma after a fall from a bicycle when she was a child, which required suture.

Figure 1: Yellowish erythematous plaques with well-delimited borders, telangiectasias and atrophic center in the breast.

Figure 2: Erythematous papular lesions on the right forearm, found during physical examination.
The patient denied pre-existing diseases, such as diabetes mellitus, and did not use any continuous medication.

After clinical evaluation, a biopsy of one of the breast lesions was performed for histopathological analysis, which revealed a superficial and deep dermal perivascular inflammatory infiltrate, composed of lymphocytes and plasma cells, with histiocytes grouped between the collagen bundles, as well as sclerosis of the reticular dermis, confirming the diagnosis of NL (Figure 3).

The use of topical tacrolimus 0.1% ointment and oral acetylsalicylic acid 100 mg/day was proposed as treatment.

Discussion

NL is an idiopathic granulomatous disease typically associated with diabetes [1]. The incidence of NL in patients with diabetes is estimated at 0.3% to 1.2%, being more frequently observed in patients with type 1 diabetes. NL precedes diabetes in up to 14%, appears simultaneously in up to 24%, and occurs after diabetes is diagnosed in 62% of cases [1]. Other conditions already associated with NL are thyroid disorders, inflammatory bowel diseases, rheumatoid arthritis and sarcoidosis. NL can also manifest in healthy individuals with no underlying disease [1]. There is a predominance of females, however, the course of the disease seems to be more severe in men, as they are more likely to develop ulcerations [1].

The pathogenesis of NL has not been completely elucidated. Vascular disorders with deposition of immune complexes, or microangiopathic alterations that lead to collagen degeneration seem to be involved in the pathophysiology of the disease. Interestingly, there seems to be a higher incidence of retinopathy and systemic microvascular disease in diabetic patients with NL. Other hypotheses include abnormalities in collagen production and dysfunction in neutrophil migration [1].

Lesions are commonly found in the lower limbs, particularly in the pre-tibial region, which is affected in 85% of cases. However, there are reports of cases with lesions on the face, genitalia, trunk, scalp and upper limbs [1].

Typical lesions start as asymptomatic erythematous papules and nodules that progress to well-defined yellowish plaques with telangiectasias and central atrophy. An erythematous border may be present in the inflammatory phases of the disease. The clinical course is variable, however most patients have a chronic course and the disease may be resistant to several therapies. Ulceration is present in up to one third of patients with NL. Squamous cell carcinoma, although a rare complication, can appear in long-lasting NL lesions, even without the presence of ulceration [1].

The treatment of NL can be challenging, and so far there are no controlled and comparative clinical trials to evaluate the best therapeutic option. Medications reported in the literature include topical, intralesional and systemic corticosteroids, Phototherapy with psoralen-UV A (PUVA) and topical tacrolimus. The best therapeutic response is observed in the early inflammatory stages, while little improvement is expected when atrophy is already established. Systemic treatments aimed at improving microcirculation and antiplatelet aggregation, such as aspirin, dipyridamole, and pentoxifylline, have also been reported but appear to have limited success as Monotherapy [1].

NL occurring on preexisting surgical scars has been reported in the literature (2-12). Sahl reported in 1978 the case of a 28-year-old diabetic woman who developed generalized NL lesions, including a group of confluent lesions over a scar from a cholecystectomy performed 8 years earlier [3]. The author attributed to structural changes in connective tissue and vascularization of scar tissue a possible greater susceptibility of scars to being sites of inflammatory, infectious, neoplastic and granulomatous diseases [3]. At least ten other cases have been reported occurring in both diabetic and non-diabetic patients [4,5]. The time interval between surgery and the onset of lesions has ranged in the literature from 6 months up to 38 years, and most cases reported occurred in middle-aged diabetic women [6].

Among the possible pathogenic mechanisms, trauma seems to serve as a trigger for the development of NL, given the propensity of NL to appear in the pre-tibial region, a site frequently exposed to physical trauma [5]. The appearance of NL lesions in pre-existing surgical scars has been interpreted by most authors as an example of Koebner’s isomorphic phenomenon [6]. Koebner’s phenomenon, which denotes the appearance of isomorphic new skin lesions of a pre-existing dermatosis on areas of cutaneous injury in otherwise healthy skin, has been described in several skin diseases, including psoriasis, vitiligo, and lichen planus [7]. Although different traumatic stimuli such as excoriations, bites, burns, laser therapy and radiotherapy have been reported in association with Koebner’s phenomenon, almost all published cases of Koebner’s phenomenon and NL occurred in...
surgically induced scars, which made Acebo, et al. suggest the need for deeper injuries in the dermis for the development of lesions [6].

In a literature review, Prieto-Torres, et al. observed that of the ten reported cases, four had a previous history of NL, while in the other cases the lesions over scars were the first manifestation of the disease, and questioned whether it was a Koebner isomorphic phenomenon or a Wolf isotopic response [4]. Wolf’s phenomenon was originally described as the occurrence of a new skin disease at the site of a previously healed herpetic eruption, although the same phenomenon has been reported subsequently in sites affected by dermatological conditions not related to herpes [4].

There are at least three cases of NL appearing on surgical scars of the breast reported in the literature [5,8,9]. Azoulay, et al. reported the case of a 55-year-old diabetic woman who initially underwent a mastectomy during treatment of breast cancer, and subsequently underwent surgical reconstruction of the breast with the placement of a silicone implant. Two years after the reconstruction, she had a prosthetic rupture and an implant replacement was performed. Four months after this prosthesis replacement, the patient developed two NL lesions over breast scars. The patient had a previous diagnosis of NL, with lesions in her left leg for over ten years [8]. Khurana, et al. reported a case of NL on a mastectomy scar of a non-diabetic woman who underwent bilateral mastectomy and breast reconstruction surgery due to breast cancer. The NL lesion appeared 6 years after surgery [5,10-12].

Our case highlights a rare presentation of NL, and reinforces the theory that trauma plays a role in the pathogenesis of this disease. In our patient, who did not have diabetes or a previous history of NL, the lesions appeared 13 years after breast augmentation surgery with silicone implants, in accordance with the large interval observed in previous reports. It is noteworthy the occurrence of simultaneous lesions in scars with different origins, locations and ages, since the patient also had a lesion on a scar from an accident in her childhood, that is, much older than surgical scars.

References