

CASE REPORT

Necrobiosis Lipoidica Diabeticorum

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Introduction: Necrobiosis lipoidica diabeticorum is a rare complication occurring mostly in diabetic patients appearing as a skin redness that soon transforms into an extensive necrosis. The usual aspect of such lesions is a deep ulceration with irregular borders developing especially on the lower limbs. **Case presentations:** This paper intends to present two cases of necrobiosis lipoidica diabeticorum focusing on the clinical aspect of this disease and the original treatment applied in the Plastic Surgery Department of the Clinical County Hospital of Targu Mures. Both patients suffered from type II insulin-requiring diabetes. The first case is a 63 year old female with different stages of necrobiosis lipoidica diabeticorum lesions developed on the anterior aspect of both legs. This patient was treated using only conservative methods. The second case is a 64 year old male who developed an extensive full-thickness necrosis on the right dorsal aspect of the hand and forearm. The lesions required conservative treatment and the surgical debridement of the extensive necrotic tissues. **Conclusions:** Necrobiosis lipoidica diabeticorum is a dramatic condition requiring a well informed approach in order to save the healthy tissues as much as possible. In both cases, the wounds healed spontaneously after a long period of time.

Keywords: necrobiosis lipoidica diabeticorum, conservative treatment, type II insulin-requiring diabetes

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Introduction

Necrobiosis lipoidica diabeticorum is a rare and chronic granulomatous skin disorder of little known etiology that affects 0,3% of diabetic patients. More than 50% of those diabetic patients require insulin treatment [1,2]. There are literature reviews mentioning cases occurring in non-diabetic patients but they suffered from other comorbidities such as essential hypertension, cardiac failure, obesity or dyslipidemia. The condition was first described in 1929 by Oppenheim [1].

Regarding its epidemiological aspect, it is three times more frequent in females than in males. The average age of onset is 30 years old, but there are cases reported in all age groups, including children [1,2,3].

The etiology is unknown, but there are several theories trying to explain the mechanism:

- Diabetic microangiopathy – the leading etiologic theory, due to the association between the disease and diabetes [4]. Similar changes can be found in diabetic nephropathy and retinopathy;
- The deposition of immunoglobulins, the third component of complement and fibrinogen in the blood vessel walls [1,4,5];
- Abnormal collagen formation and organisation [4];
- Abnormal glucose transport by fibroblasts [1];
- Inflammation, trauma and metabolic changes.

The diagnosis of necrobiosis lipoidica diabeticorum is mainly clinical. In rare cases, when the clinical exam is inconclusive, a biopsy of the lesion might be required for diagnosis.

The typical localisation for those lesions is the lower limbs (most of them on the pretibial area) and are usually multiple and bilateral, but they can also be unilateral.

Other atypical presentations involve the face, trunk, scalp and upper limbs.

Patients usually present with a characteristic asymptomatic erythema which slowly enlarges over different periods of time ranging from several days-months to one year. The size of those areas can vary from 1-3 mm to huge dimensions, are well-delimited and initially red-brown in colour and progress to yellow, shiny, atrophic plaques [4]. Ulceration at the site of minor trauma (Koebner phenomenon) is a rare complication. [6]

Pain could be one of the principal symptoms of this disease, but due to the diabetic neuropathy many lesions are painless.

Marginal biopsy of the lesion and pathology examination of the sample shows characteristic granulomas in the dermis and subcutaneous tissue disposed in a layered fashion. Between the granulomas one can find degenerated collagen, cholesterol, mucin and fibrin deposits. The main components of the granuloma are histiocytes, lymphocytes, occasional plasma cells and eosinophils [1]. There is also a decrease in the number of intradermal nervous ends, blood vessel wall thickening and endothelial cells swelling.

Direct immunofluorescence microscopy can reveal the presence of IgA, IgM, the third component of the complement or fibrinogen in the blood vessel walls [5].

Differential diagnosis can refer to dermatologic pathologies such as granuloma annulare, xanthomas, xanthogranulomas, amiloidosis and rheumatologic disorders (sarcoidosis, rheumatoid arthritis). [4]

This paper intends to present two cases of necrobiosis lipoidica diabeticorum focusing mostly on the clinical aspect of this disease and the original treatment applied in the Plastic Surgery Department of the Clinical County Hospital of Targu Mures.

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Case number 1

The first case is a 63 year old female with type II insulin-requiring diabetes, rheumatoid arthritis and operated colangiocarcinoma, who presented to our department with an extensive necrobiosis lipoidica lesion. The lesion showed a characteristic aspect – large size and well-defined borders and was located on the anterior aspect of the left leg. The onset and evolution were slow, finally reaching the aspect shown in Figure 1A.

The ulceration was treated with daily or every two days dressings (depending on the amount of exsudate), using nitrocellulose dressings. After approximately 2 months, the upper 2/3 have been debrided and a good granulation tissue appeared, while the inferior 1/3 has shown the devitalised tendon of the anterior tibialis muscle (Figure 1B).

After 3 months, the initial ulceration has been completely debrided. The granulation tissue was covered by the new epithelium, excepting a small portion of the proximal part of the wound (Figure 1C). After about 2-3 more weeks, the entire ulceration has been debrided and healed by spontaneous epithelialization, showing a shiny new epithelium, which will slowly go through the wound contraction phase (Figure 1D).

During her last appointment for follow-up treatment and consultation (Figure 1E), the aspect of the lesion has improved a lot. New scarring tissue appeared along with a slight hiperkeratosis and desquamation. The next step is the daily application of different hydrating cremes which substantially improved the aspect and the evolution of the scar tissue, stabilizing the neoeptithelium and preventing hiperkeratosis (Figure 1F).

Unfortunately, while the left leg was healing, a new ulceration appeared on the anterior aspect of the right leg with a characteristic aspect for necrobiosis lipoidica diabetorum (Figure 1G). Fortunately, this lesion was limited to the superficial skin and suprafascial subcutaneous tissue.

This wound was also treated conservatively using thick absorbent nitrocellulose dressings that were changed every time they were soaked with exsudate. In the case of this second wound, the exsudate was not so abundant and the evolution towards healing was faster (Figure 1H). Unfortunately, the patient developed multiple metastases due to the intrahepatic cholangiocarcinoma, so she couldn't come to the hospital anymore for follow-up consultation and treatment.

Case number 2

The second case is a 64 year old male with type II insulin-requiring diabetes and chronic renal failure undergoing dialysis 3 times a week. He spontaneously developed an extensive necrosis on the posterior aspect of the right forearm and right hand (Figure 2A). The necrosis involved the superficial skin and suprafascial subcutaneous tissues. It could also be interpreted as a secondary necrosis (vascular type) due to the trombosis of one of the therapeutic fistulas used for dialysis, but the evolution and clinical aspect leans more towards necrobiosis lipoidica diabetorum.

The patient was admitted in the nephrology department but he was brought daily to the plastic surgery dressing room to undergo local treatment. Initially, the necrotic tissues were surgically debrided on behalf of scissors and forceps. The excision was made at the demarcation line between the necrotic and the healthy tissues. The large defect on the posterior aspect of the arm and fingers was then covered daily by alternating silver sulfadiazine and povidone-iodine dressings which led to the progressive autolytic debridement of the slough and fibrin deposits (Figure 2B).

Two-three weeks later, the necrotic lesions were slowly replaced by granulation tissue, with adherent slough and abundant fibrin deposits still present, along with an important exsudation with its characteristic foul odour. Slowly,

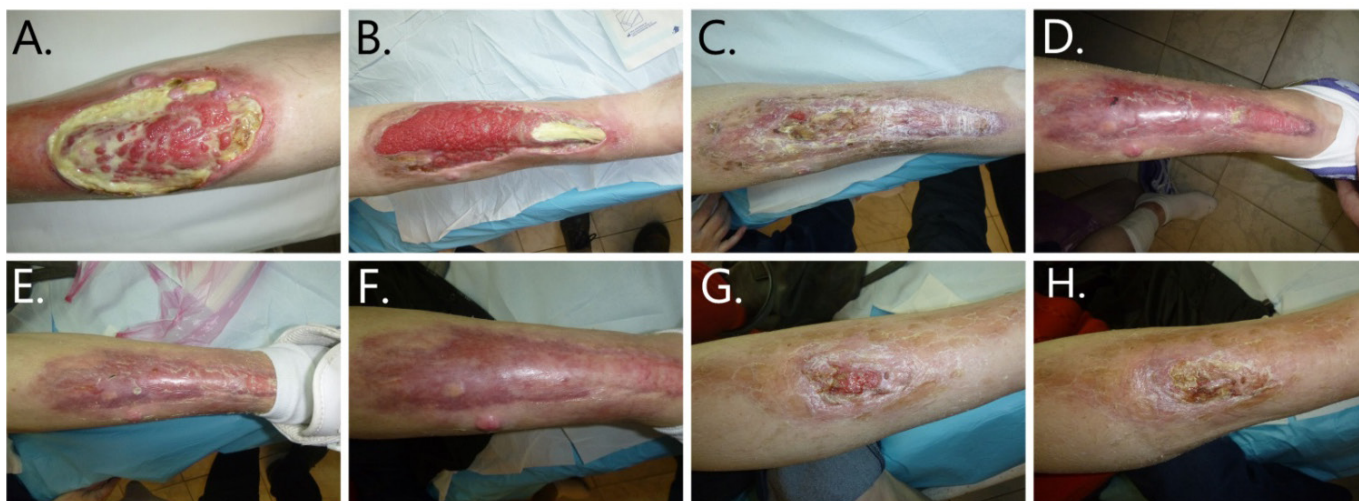


Fig. 1. Extensive necrobiosis lipoidica diabetorum lesions in a 63 year old female. One can see the initial aspect of the left leg wound (A) and different stages of evolution during the treatment period: 2 months (B), 3 months (C), 4 months (D), after the formation of the scarring tissue (E) and after the application of hydrating cremes (F). While the lesion on the left leg was healing, another one appeared on the right leg (G). The same conservative treatment was used (H).

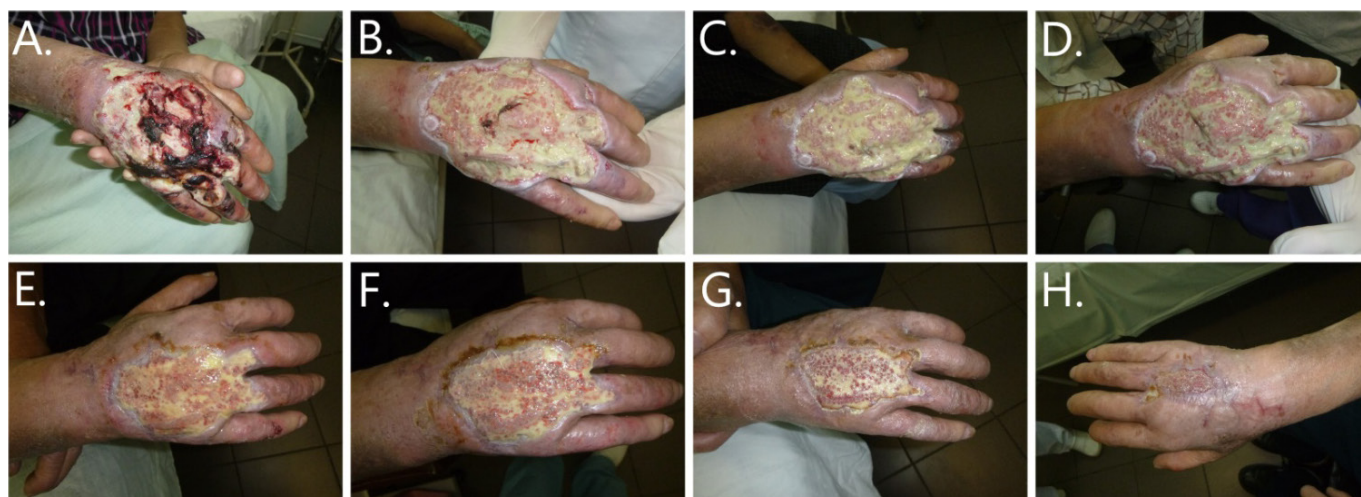


Fig. 2. Necrobiosis lipoidica diabetorum lesions in a 64 year old male, who developed a spontaneous necrosis on the right forearm and hand (A). The surgical removal of the necrotic tissues and the autolytic debridement of the wounds was performed (B). Several stages of the wound evolution during the autolytic debridement can be seen in pictures B-G, leading to the final aspect (H).

the dressings were replaced by specific polyurethane foam dressings and nitrocellulose dressings (Figure 2C).

After the application of the absorbent dressings, which encouraged the autolytic debridement process, one can see that the exsudate and fibrin deposits have significantly diminished and a granulation process started (Figure 2D).

The autolytic debridement and granulation development have been two long term processes due to the fact that deep vein thrombosis appeared on the right forearm and arm, which eventually led to the recurrence of the abundant exsudate and formation of new fibrin deposits but with a lower intensity. About 2-3 months have passed from the beginning to the wound aspect seen in Figure 2E.

In figure 2F, one can see how (when alternating the four therapeutic factors silver sulfadiazine, povidone-iodine, polyurethane foam dressings and nitrocellulose absorbent dressings), the initial lesion shrunk progressively and the granulation tissue islands increased in number.

About 4 months later, the wound margins started to get progressively smaller, the lesion reaching less than half of its initial size. A thin marginal epithelialization lining appeared which will lead to the reduction of the lesional surface (Figure 2G).

Finally, after 4-5 months of conservative treatment, the whole lesion healed completely with minimum scarring and about 70% of its initial functionality gained back. To be noted that conservative treatment was the only way to keep the right upper limb, since the already existing therapeutic fistulas and the frequent thrombosis would have been an indication for above elbow amputation. Even though the therapeutic period was long, the treatment was a success, since the final morphological and functional result was spectacular, saving the patient's hand (Figure 2H).

Discussions

In necrobiosis lipoidica diabetorum, the surgical removal of slough and necrotic tissues is not suitable for the wound healing process because such aggressive procedures usually remove healthy tissues and new formed blood vessels which are extremely important for the proliferative phase of healing. So, we preferred to deal with it conservatively, excepting the full-thickness necrosis located on the distal part of the right upper limb as mentioned above.

Conclusions

Necrobiosis lipoidica diabetorum is a dramatic condition, requiring a fully documented surgical approach in order to prevent unnecessary loss of healthy tissues. All cases treated in the last 20 years in the Plastic Surgery Department of the Clinical County hospital of Targu Mures healed spontaneously in a pretty long period of time but with no need for skin grafting of the extensive ulcerations and almost no need for mutilating amputations.

Authors' contribution

C.A. (Data collection and analyzing; Conceptualization; Writing – original draft)

B.A. (Data collection; Study coordinator; Conceptualization; Writing – supervision, review & editing)

Conflicts of interest

None to declare.

Informed consent

This paper is one of the chapters of my graduation degree for which data collection was approved by the Ethics Committee.

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