

# Necrobiosis lipoidica: A clinicopathological study in the Indian scenario

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## ABSTRACT

**Context:** Necrobiosis lipoidica (NL) is a chronic granulomatous dermatitis that is commonly associated with diabetes mellitus. Most of the current knowledge about this entity is from western literature. **Aims:** This study evaluates the clinicohistological features of NL in an Indian scenario. **Materials and Methods:** We retrospectively reviewed clinical features, associated comorbidities, and biopsies of all patients with NL over a period of one year. **Results:** Five cases of NL were seen during the duration of the study. The preliminary clinical diagnosis ranged from sarcoidosis to tinea incognita. The commonest clinical presentation in the Indian scenario was of asymptomatic erythematous to skin-colored plaques and nodules on the shins with or without central atrophy. The most common site of involvement was the shin (3 of 5 patients). NL was associated with Diabetes mellitus in only two cases, both of whom were male patients. On histology, various patterns of inflammation were seen including the palisading, interstitial, and mixed granulomatous infiltrates. One patient had sarcoidal granulomas in association with an interstitial pattern. Features seen consistently in all cases include perivascular lymphoplasmacytic infiltrates, interstitial lymphocytic infiltrates, and fibroplasia. Interstitial mucin deposition was not observed in any of the biopsies. **Conclusion:** The diagnosis of NL was missed in most cases due to the rarity of the disease, absence of concomitant diabetes, and atypical presentations. Histology was a useful tool in clinching the diagnosis.

**Key words:** Diabetes, necrobiosis lipoidica, palisading granuloma

## INTRODUCTION

Necrobiosis lipoidica (NL) is a chronic granulomatous dermatitis that is commonly associated with diabetes mellitus.<sup>[1]</sup> Most of the current knowledge about this entity is from western literature. In the Caucasian population, it typically presents as yellow brown telangiectatic sclerodermiform plaques on the shins with central atrophy and raised violaceous borders. This classical presentation is not always seen; hence, the diagnosis is usually based on clinicopathological correlation.<sup>[2]</sup> There is very limited literature evaluating NL in an Indian scenario as it is relatively rare in our population.<sup>[3,4]</sup> We studied five patients with NL who presented to a tertiary care hospital in western India. The clinical and histological features that deviate from the usual pattern, interesting and easily misleading findings are described.

## MATERIALS AND METHODS

We retrospectively reviewed clinical features and associated comorbidities of all patients with

NL whose biopsy samples were received at a dermatopathology referral service in western India. The study was conducted over a period of one year (January 2011-December 2011). Hematoxylin and eosin (H and E)-stained sections of their skin biopsies were also reviewed for clinicopathological correlation.

## RESULTS

During the period of the study, 5 600 biopsy samples from various parts of the country had been received by our center, five of which were consistent with NL.

### Age and gender

The male to female ratio was 2 : 3. The mean age of the patients was 51 years (range, 29-64 years). The mean age of the male patients was 59 years, and that of the female patients was 37 years. The clinical profile of the patients is summarized in Table 1.

### Preliminary clinical diagnosis

Lesions were present unilaterally in three of the five patients. The commonest clinical

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**Table 1: Clinical profile of Necrobiosis Lipoidica**

	Case 1	Case 2	Case 3	Case 4	Case 5
Age (yrs)	29	30	64	48	54
Sex	Female	Female	Male	Female	Male
Diabetic status	Non diabetic	Non diabetic	Diabetic	Non diabetic	Diabetic
Type and color of lesions	Skin-colored asymptomatic plaque	Erythematous plaque	Erythematous nodule	Skin-colored asymptomatic plaque	Yellowish asymptomatic plaque
Clinical diagnosis	Sarcoidosis, granuloma annulare	Dermatophytosis, granuloma annulare	Erythema nodosum	Sarcoidosis, granuloma annulare	Necrobiosis lipoidica, myxedema



**Figure 1:** Necrobiosis Lipoidica in Indian Skin: asymptomatic erythematous to brown-colored plaques with central atrophy

presentation in the Indian scenario was of asymptomatic erythematous to skin-colored plaques and nodules on the shins with or without central atrophy [Figure 1]. The characteristic yellowish brown plaque with a peripheral violaceous hue was observed only in one patient who was an elderly diabetic [Figure 2]. Telangiectasias were not visualized in any of our five patients. Central atrophy was observed in three of the five patients. The most common site of involvement was the shin (3 of 5 patients). NL was associated with Diabetes mellitus in only two cases, both of whom were male patients. The preliminary clinical diagnosis ranged from sarcoidosis to tinea incognita. Other diagnoses that were considered included cutaneous vasculitis, granuloma annulare (GA), and myxedema. A differential diagnosis of NL was considered in only one patient, who presented with typical clinical features as described earlier. He had concomitant diabetes mellitus. Cutaneous anesthesia was not present in any of our patients.

### Histopathological features

The epidermis was normal in all except one case in which we observed a hyperkeratosis with spongiosis. Transepidermal elimination and ulceration were not observed in any of our five cases. Dilatation of the papillary dermal capillary network was seen in 3 of 5 biopsies. The entire dermis was involved in all cases. Various patterns of dermal inflammation were observed in our case series including palisading [Figure 3]

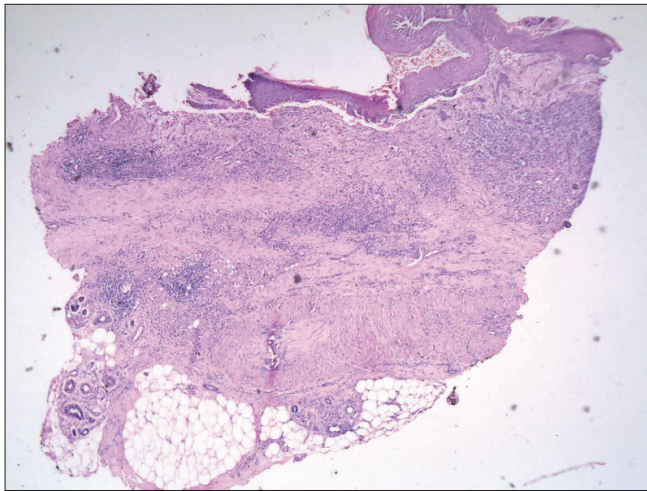


**Figure 2:** Classical Necrobiosis Lipoidica: yellowish brown plaque with a peripheral violaceous hue

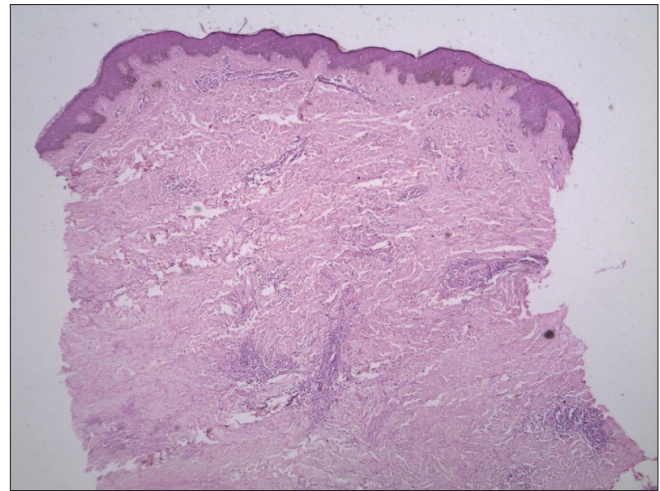
interstitial and mixed (palisading + interstitial) granulomatous infiltrates [Figure 4]. One patient also had sarcoidal granulomas in association with an interstitial pattern [Table 2]. The classical “tiered” or “wedding cake” was seen in only one case. Giant cells were seen in all five biopsies of which four had Langhans type and one had foreign body type giant cells. Other consistent features found in all cases included perivascular lymphoplasmacytic infiltrates, interstitial lymphocytic infiltrates, and fibroplasia. Interstitial mucin deposition was not observed in any of the biopsies.

### DISCUSSION

Necrobiosis lipoidica, first described by Oppenheim under the title of dermatitis atrophicans lipoides diabetica, and later by Urbach as NL diabetorum,<sup>[4]</sup> is a granulomatous disease



**Figure 3:** Palisading granulomas in a tired/wedding cake appearance [H and E, x4]



**Figure 4:** Mixed (palisading + interstitial) infiltrate in Necrobiosis Lipoidica [H and E, x4]

**Table 2: Histopathological Features of Necrobiosis Lipoidica**

Histopathological features	No. of cases
Granuloma	5 (1 Palisading, 2 Interstitial, 1 Mixed, 1 Sarcoidal+ Interstitial)
Perivascular infiltrate (mixed)	5
Interstitial infiltrate	5
Fibroplasia	5
Telangiectasia	4
Fibrin	5
Giant cells	5 (4 Langhans type, 1 foreign body type)

of unknown etiology and pathogenesis.<sup>[5]</sup> It is thought to be associated with diabetes mellitus in 75% of cases.<sup>[6]</sup> The etiology of NL is still obscure. Focal degeneration of collagen is considered to be the initiating event. Due to its strong relationship with diabetes, many studies have focused the deposition of a glycoprotein material as in diabetic microangiopathy. Other theories implicate an antibody-mediated vasculitis which may initiate the blood vessel changes and subsequent necrobiosis in NL. Perivascular immunoglobulin deposits suggest that NL may be an immune complex vasculitis, except that the vasculitis is not demonstrable on biopsy specimens.<sup>[7,8]</sup>

NL is uncommon in India and previous reports from India do not elaborate the clinicopathological findings. In our study, none of the female patients had concomitant diabetes mellitus and their mean age was in the third decade as compared to the males who were all diabetic and with age ranging from 5-6 decade. These findings are in sync with previous studies that have found 85% of the non-diabetic patients who develop NL are women.<sup>[9]</sup>

The commonest site affected was the shins as described in classical NL; yet, due to the atypical presentations in Indians

in comparison to the classical descriptions from Western studies, the clinical diagnosis of NL was made only in one patient. This patient was an elderly male with concomitant diabetes with classical lesions. In the Indian population, the lesions did not have the classical appearance as described in western literature and commonly presents as erythematous plaques. The lesions in all the other patients mimicked various common dermatological disorders. Absence of associated diabetes as against the common belief and the atypical clinical presentations may misguide the clinician on many an occasion.

Three histopathological patterns of NL have been described in literature: the palisading granulomatous pattern, the tuberculoid pattern, and the intermediate pattern.<sup>[2]</sup> Rarely, transepidermal elimination and lymphoid follicle formation have been seen.<sup>[10,11]</sup> In our series, we found the interstitial granulomatous pattern to be the commonest. A palisading granuloma with a tired appearance which is typically described in NL was seen in only one patient. One of our patients had sarcoidal granulomas in association with the interstitial infiltrate. This is an uncommon histological pattern.<sup>[12]</sup> The clue to diagnosis was the associated necrobiosis which is absent in sarcoidosis.

There was a clinical suspicion of GA in three cases. It is also commonly associated with diabetes mellitus and can present with asymptomatic skin-colored to erythematous annular plaques primarily distributed on the extremities. Histopathologically, lesions of NL can be differentiated from GA by the presence of plasma cells, perivascular fibrin deposition, and the relatively lower dermal mucin deposition.<sup>[13]</sup>

Sarcoidosis is another close differential diagnosis and differs from NL by the absence of necrobiosis. The most consistent histopathological features of NL found in our study include interstitial granulomas, lymphoplasmacytic infiltrate, and telangiectasia.<sup>[14]</sup>

## CONCLUSION

The diagnosis of NL was missed in most cases due to the rarity of the disease, absence of concomitant diabetes, and atypical presentations. Histology was a useful tool in clinching the diagnosis. In this study, done over one year, we found five cases of NL. Further studies are required to determine if the incidence of NL is on a rise in the Indian population.

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