Necrobiosis Lipoidica

Necrobiosis lipoidica is an inflammatory skin disorder characterised by irregularly shaped, callous lesions with reddish-brown pigmentation and central atrophy. It was originally described in conjunction with diabetes - in 1929, by Oppenheim who coined the name *dermattitis atrophicans lipoidica diabeticca* and, in 1932, by Urbach who called it *necrobiosis lipoidica diabetica*. As it occurs in the absence of diabetes as well, it is now known as necrobiosis lipoidica, a term applied to all cases, whether occurring in those with diabetes or not.

The pathology is collagen degeneration with granulomatous response, associated with thickened blood vessels and fat deposition. The underlying cause is unknown but is believed to involve microangiopathy.\(^1\)

Epidemiology\(^2\)

It is a rare skin condition. Whilst there is a high prevalence of diabetes mellitus in patients with necrobiosis lipoidica (one third of cases have diabetes, and two thirds have glucose tolerance abnormalities), the reported prevalence of necrobiosis lipoidica in patients with diabetes is 1-2%. It most commonly presents in the 30s but can present at any age, including infancy. It tends to appear earlier in those with diabetes than in others: in one study, approximately 2% of young people with diabetes (aged up to 22 years) had a necrobiosis lipoidica lesion compared with none of the control subjects.\(^3\) It is three times as common in women as it is in men. Non-diabetic familial clustering of necrobiosis lipoidica does occur but extremely rarely.

Smoking is more prevalent in patients with diabetes with necrobiosis lipoidica than in those without it, as are diabetic complications (retinopathy and nephropathy), although the presence of necrobiosis lipoidica does not correlate with diabetic control.

Presentation

- Shiny patches slowly enlarge over months or years. They are initially a reddish brown and 1-3 mm in diameter but progress to yellow and become depressed and atrophic plaques.
- The most common site is the pretibial area but they can occur on the face, scalp, trunk and upper arms where they are less likely to be correctly diagnosed.
- There is often no pain (due to associated neuropathy) but it can be very painful.
- Trauma produces ulceration.
- Köbner’s phenomenon may be demonstrated, in which lesions occur in areas of trauma. (This phenomenon is more typically associated with psoriasis and lichen planus.)
Differential diagnosis

Usually the appearance is fairly typical but variations can be difficult to diagnose. Consider as a cause of atypical leg ulcers in diabetic patients.\[4\]

- Superficial annular lesions may look like granuloma annulare. However, granuloma annulare do not exhibit the typical yellow fatty appearance of necrobiosis lipoidica plaques.
- Yellow, fatty lesions may resemble xanthoma.
- Sarcoidosis can appear very similar, even on histology.
- Erythema nodosum. These lesions do not ulcerate.
- Rheumatoid nodules are similar histologically but tend to be raised rather than atrophic. Ulcerated necrobiotic areas have been described in rheumatoid arthritis.
- Varicose eczema produces a scaly rash and is usually near the malleoli.

Investigations

If the patient is not known to have diabetes this must be checked. Biopsy of the lesion may be helpful but be aware of poor wound healing.

Management\[2, 5\]

Management is impaired by lack of understanding of the aetiology of the condition. No treatment to date is completely effective and, whilst numerous treatments have been tried, none has proven effectiveness based on controlled trials.

- Trauma should be avoided, and strategies for prevention of ulcers employed. Wound care for established ulcers is as for other diabetic ulcers.
- Potent topical steroids are usually considered first-line treatment. This may reduce inflammation but it does not benefit burned-out lesions and may aggravate atrophy, so careful monitoring for this is required. Intralesional injections of steroids are also sometimes helpful, but increase the risk of ulceration.
- Immunomodulating drugs have also been used, with varying levels of success, to treat necrobiosis lipoidica:
  - Ciclosporin\[6\]
  - Topical tacrolimus\[7\]
  - Anti-tumour necrosis factor alpha (anti-TNF-α) therapies\[8\]

Studies demonstrate spontaneous healing of necrobiosis lipoidica following pancreas and kidney transplantation and the immunosuppressive regime is though to have played a significant role in this.\[9, 10\]

- Antiplatelet treatment seems logical but controlled trials have had different results. Aspirin and dipyridamole have been used. Pentoxifylline decreases blood viscosity and increases fibrinolysis and erythrocyte deformity and it may be helpful. Ticlopidine and perilesional injections of heparin have been used in uncontrolled trials.
- Excision and grafting are occasionally used but poor healing and recurrence are common.
- Phototherapy. Photodynamic therapy has been used, as have topical retinoids and topical psoralens with ultraviolet A (PUVA).
- Laser treatment has been used to stabilise lesions and reduce erythema and telangiectasias.

Prognosis

The lesions do not heal well and are usually considered a chronic, relapsing condition. They are known to remit spontaneously and even resolve. The most common complication is ulceration, but occasionally squamous cell carcinoma can arise in areas of long-standing necrobiosis lipoidica.\[1, 11\]
Further reading & references

- Necrobiosis lipoidica; DermNet NZ
- Related conditions - Necrobiosis lipoidica; Diabetes UK


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