2. Necrobiosis lipoidica

Necrobiosis lipoidica (NL) is a chronic skin condition associated with dermal collagen degeneration, decreased collagen synthesis, granulomatous infiltrate, atrophy, and lipid deposition. Because NL frequently presents in diabetic or glucose-intolerant individuals, it is often referred to as necrobiosis lipoidica diabeticorum. Conversely, only about 0.7% of diabetic patients have NL. It is twice as common in women as it is in men.\(^1\)

Single or multiple lesions usually present in the pretibial area, initially as small red-brown erythematous markings that enlarge over time. Due to granulomatous inflammation and degeneration of collagen, combined with atrophy, the lesions evolve into waxy yellowish-red plaques with depressed centres. The shiny, telangiectatic surface has been described as having a “glazed-porcelain” sheen. The border is often raised and red-violet in colour. In one-third of cases, lesions develop ulcerations that heal poorly.\(^2\)

Necrobiosis lipoidica is generally painless and patients mainly complain about aesthetic issues. The ulcerations within the NL lesions, on the other hand, usually present with pain. As this condition is chronic, a typical patient will endure several cycles of NL lesion growth and stagnation, and the course of each cycle is unpredictable.

The pathogenesis of NL is largely enigmatic. It has been suggested that the cause of NL is microangiopathic, owing to the clear association with diabetes. In fact, it has been shown that NL in diabetic individuals might be a warning sign for diabetic nephropathy and retinopathy;\(^3\) whether or not poor glucose control is correlated with the development and progression of NL lesions remains controversial.\(^4\) Trauma seems to play a role in the pathogenesis, as evidenced by the isomorphic response (Köbner phenomenon), as lesions develop on sites of minor tibial injury or other areas of the body after surgical trauma. A defect in collagen and elastin remodeling in affected patients has also been proposed as a contributing factor.

Diagnosis

Diagnosis of NL is usually made clinically, but a biopsy might be necessary in atypical lesions. In the case presented, the pretibial location of the lesion, its chronic nature, and the diabetic status of the patient are all very suggestive of NL. When it presents atypically, however, NL can be difficult to differentiate from other similar disorders. This is especially true in early stages of NL, when it cannot yet be established as a chronic condition. In an atypical case, a biopsy can provide confirmation of NL, although a good examination and a history of the lesion might help to rule out, or at least dampen the probability of, other similar diseases. It should be noted that although granuloma annulare lesions lack the typical fatty appearance of the yellow plaques of NL, early lesions of NL can easily be mistaken for granuloma annulare and the histology can be confusingly similar. Necrobiotic xanthogranuloma is very rare, and can resemble NL because of its yellow-red nodules or plaques; however, in contrast to NL, necrobiotic xanthogranuloma typically presents periorbitally and is strongly associated with paraproteinemia. Sarcoidosis is a systemic granulomatous disease that affects not only the skin but also multiple organ systems; it is largely a diagnosis of exclusion. Although the hallmark of sarcoidosis is a granulomatous plaque, cutaneous lesions in sarcoidosis very rarely atrophy or ulcerate. Erythema nodosum presents with bilateral, tender, erythematous nodules that lack epidermal change, atrophy, or ulceration.

Treatment

Treatment of NL is often disappointing and spontaneous resolution is unusual. The main objective of NL treatment is to halt the progress of developing lesions. Smoking cessation should be advocated. Topical or intralesional corticosteroids are first-line therapy for new or developing lesions, while oral corticosteroids or even dermal skin grafting might be indicated in severe cases. Laser treatment, topical psoralen with ultraviolet A, and topical calcineurin inhibitors have also been described as therapeutic modalities. Strict avoidance of trauma is suggested to prevent ulceration and the development of new lesions.

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Competing interests

None declared

References