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Necrobiosis lipoidica

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ABSTRACT

Necrobiosis lipoidica is a rare granulomatous dermatosis, we report observation a of 17-year-old patient, with any pathological history, has presented erythematous oval plaques with an atrophic yellowish center at the anterior surfaces of legs. The diagnosis of necrobiosis lipoidica was confirmed after biopsy. In view of this situation, additional tests were requested. They revealed an unknown diabetes. The anti-GAD antibodies and anti-IA2 antibodies were positive and the patient was placed on insulin therapy. The evolution was marked by a spontaneous regression of lesions.

Keywords: Necrobiosis lipoidica, diabetes

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INTRODUCTION

Necrobiosis lipoidica (or Oppenheim-Urbach disease) is a rare granulomatous dermatosis. It occurs in an average of 0.3 to 1.2% among diabetics [1]. This dermatosis is located generally on the legs. The lesions appear as well-circumscribed erythematous plaques, with central depressions. No treatment is specific to this disease

Observation:

A 17-year-old patient, with any pathological history, has presented erythematous oval plaques with an atrophic yellowish center at the anterior surfaces of legs. Those lesions were symmetrical and asymptomatic (Figure 1, 2).

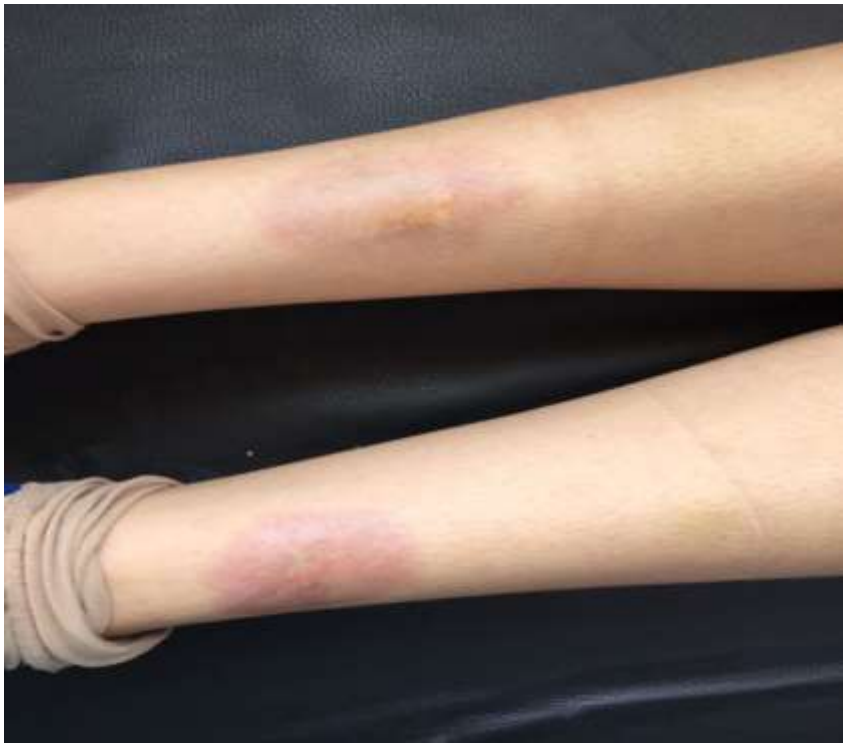

Figure 1: Lesions were symmetrical

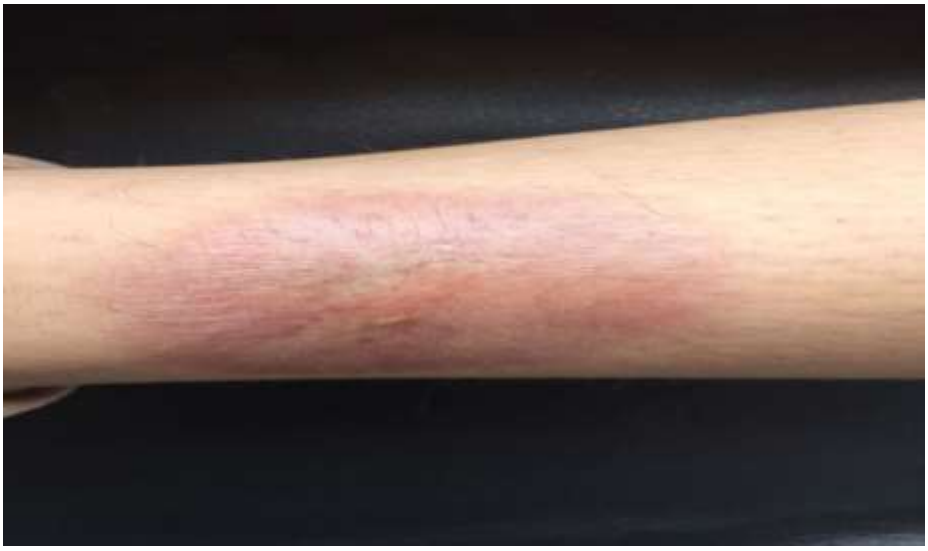

Figure 2: Lesions were asymptomatic

The diagnosis of necrobiosis lipoidica was raised and a biopsy was performed. It showed a nodular granulomatous inflammatory reaction with a palissadic border arranged around the poorly defined foci of altered connective stroma. A giant Plurinucleated cells and dendrocytes were present. A hyaline aspect and lipid deposits were also recognized. In view of this situation, additional tests were requested. They revealed an unknown diabetes, with fasting blood glucose at 1.45 g / l and glycated hemoglobin at 8.4%. The anti-GAD antibodies and anti-IA2 antibodies were positive and the patient was placed on insulin therapy. The evolution was marked by a spontaneous regression of lesions

DISCUSSION:

Necrobiosis lipoidica, formerly known as the Oppenheim-Urbach disease, is an inflammatory granulomatous dermatosis which was first described by Oppenheim in 1929. In 1932, Urbach proposed the name "Necrobiosis lipoidica diabeticurum "[2].

It's reported in 0.3 to 1.2% of diabetics, with a predominance of the female side, which starts in the third or fourth decade of the age [3]. The association of Necrobiosis lipoidica with diabetes is commonly accepted, but controversial. According to studies, it varies from 11 to 65%. However, some family cases have been described even without any association with diabetes [4].

Clinically, the lesions start with small erythematous papules that slowly grow and join each other to constitute a wide surface. They advance in the form of well-defined macules with a purplish border and a yellowish-brownish atrophic center dotted with telangiectasias [5]. It is most often located in the anterior face of the legs but can also reach the feet, the face and the upper limbs. Actually, the physiopathology of necrobiosis lipoidica remains poorly understood [5]. Vascular changes associated with antibody deposition may be involved in the development of the disease [2].

The diagnostic confirmation is histological. It shows a granulomatous infiltrate over the entire thickness of the dermis. It is placed in a palisade around the rearranged foci of connective tissue. The infiltrate includes lymphocytes, positive Factor XIIIa dendrocytes, histiocytes, plasmocytes as well as epithelioid cells and plurinucleated giant cells. We can also find a pseudo-tuberculoid aspect consisting of epithelioid histiocytes with an increased number of lymphocytes, plasmocytes and giant cells. In immun $=histochemistry, there is an important density of dendrocytes Factor XIIIa^{+} on the entire surface of the lesion [2]. The evolution is most often chronic. Even if spontaneous regression is observed in 20% of the cases (as was the case of the observation reported), the situation can be complicated by ulceration that appears in 35% of the cases [3]. The degeneration in squamous cell carcinoma is exceptional [6]. The balance of diabetes has no impact on the course of the disease.

If we face a necrobiosis lipoidica, it is wise to seek unknown diabetes and be vigilant regarding its eventual occurrence [7]. In the reported case, necrobiosis lipoidica has revealed diabetes.

First-line treatment is based on local or intralesional corticosteroids. Therapeutic alternatives include tacrolimus topical[8], PUVA therapy, dynamic phototherapy[9]. The immunosuppressants and anti-TNFα can be used in more severe cases [10]. Actually, the therapeutic is still a challenge while waiting for a better pathobiological knowledge.

CONCLUSION:

Usually benign, Necrobiosis lipoidica is a rare affection. Its association with diabetes is not exceptional, but Its treatment is poorly codified and its prognosis remains a chronic dermatitis.

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