A Picture Says a Thousand Words

An uncommon initial presentation of a common disease

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CASE PRESENTATION

A 10-year-old female of First Nations heritage presented with a 1-year history of worsening and enlarging slightly pruritic skin lesions on both her shins. The lesions were previously diagnosed and managed as eczema and treated with topical corticosteroids with no effect. Upon follow-up with her primary care physician, there was a 2-month history of polyuria, polydipsia, nocturia, and weight loss. Her weight was 32 kg, height was 144.8 cm, and body mass index was 15.3 kg/m². Serum glucose was 34.5 mmol/L and HbA1c was 14%. She was not in diabetic ketoacidosis. Glutamic acid decarboxylase, islet antigen, and insulin antibodies were negative. There was a family history of type 2 diabetes.

Skin lesions were present on both shins. The left lesion measured 11 × 4 cm and the right measured 8 × 3 cm. The lesions were large shiny atrophic plaques with yellow-brown colouration and some telangiectases. There was a small depression on the left medial shin thought to be consistent with a previous resolved ulceration (Figure 1).

Figure 1. Patient's left and right shins.

CASE DIAGNOSIS

The patient's recent symptoms and biochemistry were consistent with a diagnosis of type 2 diabetes. The established skin lesions were characteristic of Necrobiosis Lipoidica, a chronic granulomatous dermatitis of unknown cause (1).

Necrobiosis Lipoidica is only found in 0.5% of patients with diabetes but when present, it is a specific sign of diabetes 80% of the time and referred to as Necrobiosis Lipoidica Diabeticorum (NLD). NLD is more common in patients with known established diabetes than new onset diabetes. It is also more commonly found in adults, females, and those with type 1 diabetes compared with type 2 diabetes (2–4). In contrast, our patient was a child with newly diagnosed type 2 diabetes. The patient's extremely elevated HbA1c indicates a significant and likely prolonged unrecognized history of hyperglycemia. The extent of the NLD skin lesions is thought to not likely be associated with the degree of hyperglycemia. Prompt recognition of NLD may have resulted in an earlier diagnosis of diabetes for our patient. The natural history is usually of small plaques, often on the shins, that grow and coalesce to form larger plaques. Diagnosis is generally clinical and a biopsy is usually not required. It is reported that 19% of cases of NLD can resolve spontaneously but often over many years (5). Ulceration due to trauma is a common complication with NLD and care should be taken to avoid this as it increases the risk of infection and less commonly squamous cell carcinoma (6). There is an increased incidence of squamous cell carcinoma in NLD, and in particular more so in lesions with chronic ulceration. Dermatologically, lesions are a challenge to successfully treat. Possible treatment

includes topical glucocorticoids, tacrolimus, and tretinoin, intralesional triamcinolone, and systemic hydroxychloroquine. The successful use of TNF alpha inhibitors has also been reported (2–4). Our patient was treated with moisturizers and 0.03% tacrolimus ointment (Protopic) with limited clinical improvement.

NLD is a dermatological condition which is highly associated and specific with diabetes. Patients that present with findings suggestive of NLD should undergo prompt evaluation for type 1 or 2 diabetes.

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