An unusual skin rash in a person with type 1 diabetes

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A 73 year old man with long-standing type 1 diabetes attends for annual review. He has a previous medical history of Graves' disease treated with radio-iodine and is now taking thyroxine. He complains of a non-itchy rash on the anterior aspect of his lower legs that has developed over the past six months. Examination reveals bilateral non-pitting dermal oedema, as shown in the image of his right leg.

Questions:

1. What is the diagnosis?
2. What is the pathophysiology of this condition?

3. What treatment is recommended for this condition?

Answers

1. The diagnosis is pretibial myxoedema. This is identifiable by the characteristic pretibial site, faintly pigmented thickening of the skin, scattered papules/nodules and the bilateral non-pitting dermal oedema (1-2). Pretibial myxoedema can occasionally involve the ankles, knees or feet and typically appears insidiously over three to twelve months. It is most often asymptomatic or itchy but is sometimes painful (1-2). Pretibial myxoedema is almost exclusively seen in patients with Graves' disease and the related ophthalmopathy. It is infrequently seen in patients with Hashimoto's thyroiditis and is extremely rarely (case reports) seen in patients with no prior history of thyroid disease (3).

2. Pretibial myxoedema is the result of an inflammatory process. Cytokine-induced activation of fibroblasts leads to the secretion of acidic glycosaminoglycans (GAGs) into the dermis (4). These GAGs are hydrophilic, compress dermal lymphatics and degrade dermal collagen fibres resulting in the clinical features described above (4). Almost all individuals with pretibial myxoedema have high levels of thyroid stimulating hormone receptor antibodies, and it is presumed these activate TSH receptors on fibroblasts, the initiating factor in this cellular process (5).

3. If mild, no treatment is needed. For moderate and severe cases, first line therapy is topical corticosteroids (mid to high potency) with occlusion (e.g. wrapping legs in 'cling film') (6). Second line treatment is systemic oral steroids or steroid injection into the lesions (7). In more refractory cases, studies using immunomodulating therapies are promising; these include pentoxifylline, rituximab, plasmapheresis and intravenous immunoglobulins.

Learning Points

Pretibial myxoedema is an uncommon immune-mediated condition most commonly associated with Graves' disease with ophthalmopathy.

If symptomatic, treatment involves topical or systemic corticosteroids and if refractory, immunomodulating therapies can be tried.

References

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