Necrobiosis Lipoidica: An atypical presentation on the scalp

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Introduction

Necrobiosis lipoidica (NL) is a type of granulomatous dermatitis that classically appears on the lower extremities, particularly the perifacial surface. However, a few case reports have identified atypical presentation of lesions on the face, penis, trunk, scalp, and upper extremities. The disease has once been called Necrobiosis Lipoidica Diabeticorum as it was thought to be associated with diabetes. NL, by itself is the preferred term as only 0.01%-1.2% of patients with diabetes have NL.1 When NL is presented in an unusual location, other disease entity such as necrobiotic xanthogranuloma (NXG) should be considered. Histopathologically, NL can look similar to NXG, which also falls into a category of granulomatous reaction pattern. We review the pathologic similarities of these two conditions and discuss treatments for both types of granulomatous dermatitis.

Case report

An 85-year-old Caucasian male presents with a three-month history of nodular lesions on his left frontal scalp. The lesions are completely asymptomatic. His past medical history is significant for rheumatoid arthritis, asthma, irregular heart rhythm, and TIA. His medications include Prilosec, Lasix, Fosamax, synthroid, aspirin, hydrocodone, and Tylenol. Physical exam reveals a group of firm, skin-colored nodules about 1 cm in size without epidermal disruption on the left frontal scalp. A 3mm punch biopsy was performed. Histopathology revealed atypical necrobiotic lipoidica with evidence of a kappa light chain restricted atypical plasma cell infiltrate. This finding favors a parapneumonic necrobiotic lipoidica tissue reaction in the setting of an underlying plasma cell dyscrasia. The case was sent for a consultation to Dr. Cynthia Magro at Cornell who recommended a bone marrow biopsy and serum protein electrophoresis (SPEP) for further assessment.

On follow up, patient underwent an SPEP, which reveals no abnormality. He was subsequently referred to an oncologist for further evaluation. Since the biopsy was done, the nodule on the scalp has not grown and no new symptoms were noted. As a result, he decided not to pursue additional testing.

Clinical Photos

Rectangular punch biopsy showing layers of palisading granulomas interposed with degenerated pale collagen

Histopathology

The cellular area is infiltrated with numerous plasma cells that are well-differentiated

Discussion

Necrobiosis lipoidica (NL) is a rare granulomatous disease. The cause and pathogenesis is not well understood, but many theories have been presented. The most commonly proposed theory involves vascular disturbance with immune complex deposition or microangiopathic changes, which contributes to the development of collagen degeneration and subsequent dermal inflammation.2,3 The vascular abnormalities seen in NL are thickening of the vessel walls, fibrosis, and endothelial proliferation leading to occlusion in the deeper dermis. These characteristics were more prominent in diabetic patients than non-diabetic.4 Age of onset is typically around the third decade of life in patients with type 1 diabetes and fourth decade in patients with type 2 diabetes and in nondiabetics.5 The disease affects more females than males at a 3:1 ratio.6 Clinically, the lesion begins as multiple, small, firm, red-brown papules that gradually enlarge and coalesce into plaque. Over time these plaques become atrophic and develop central telangiectasias.7 Although the majority of lesions are painless as a result of associated nerve damage, some lesions can be painful. Ulceration can occur in up to 35% of cases following minor trauma.8 In addition, there have been reports of squamous cell carcinoma developing within long-standing NL plaques.9 NL can have similar appearance to other skin disease, particularly when the lesions occur on less common body sites. We consider necrobiotic xanthogranuloma (NXG) on our differential because of the location of the lesion and the numerous rapid plasma reagin (RPR) plasmas. NXG typically affects the periorbital area (>80%) but can also occur on the trunk and extremities. The typical presentation consists of multiple, indurated, yellow-brown plaques or nodules.10 A hallmark feature of NXG is the associated IgG monoclonal gammapathy found in 80% of patients. The most common type of paraproteinemia in IgG-kappa monoclonal gammapathy with IgG-lambda as the runner up. Patients with MGUS and NXG may have an increased risk of developing multiple myeloma and need to be monitored closely. NXG is locally destructive and can affect multiple organ systems.11 The diagnosis of NXG should prompt a thorough workup for hematologic and lymphoproliferative malignant condition, which typically manifest approximately 2.4 years after development of the skin lesions.

Histologically, both entities display full thickness involvement extending down to the subcutis. NL is described as looking like layers on a cake, having horizontal palisading granulomas interposed with degenerated pale collagen.12 Plasma cells can be seen in both entities as well as the presence of lymphoid nodules. NXG generally has broader zones of necrobiotic with granulomatous foci composed of histiocytes, foam cells, and multinucleated giant cells.13 It is more cellular with prominent stromal and foreign body giant cells. In addition, NXG will commonly display cholesterol clefts in the area of necrobiosis.

NL is difficult to treat, and there are no established treatment regimens. A multitude of case reports have described the use of several treatments, which include topical and intraluminal steroids, calcium inhibitors, intralesional interferons, antiinflammatory, antimalarials, cyclosporine, TNFα, hyperbaric oxygen, and co2 laser.14 First line treatment is topical and intraluminal corticosteroids. The use of corticosteroids should be monitored in diabetic patients especially when used on a large surface area to prevent glucose dysregulation. It should be applied to the active borders of the lesions and not to the atrophic area as this may possibly worsen the atrophy.15 Similar to NL, NXG is also difficult to treat. Some recommendations include topical, lesional, or systemic corticosteroids. Chlorambucil, cyclophosphamide, interferon alpha, antibiotics, thalidomide, plasmapheresis, and IVIG have also been tried with some success.16 Surgery is not recommended, as there is a high recurrence rate.2 In conclusion, this case demonstrates an unusual presentation of necrobiotic lipoidica on the scalp in the setting of plasma cell dyscrasia. NL and NXG may, perhaps, represent a spectrum of related disease. Therefore, it is important to give a full workup with patients in this type of scenario in case of underlying disease or systemic involvement as there are overlapping histopathologic characteristics.

References