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cutaneous lymphoid hyperplasia-grey case

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Introduction & Objectives:

Cutaneous lymphoid hyperplasia is a benign reactive process characterized by cutaneous polyclonal lymphoproliferative infiltrate. CLH presents as nodular skin lesions, mainly on the face and upper trunk. CLH may occur in response to a stimulus, often unrecognized. Known stimuli include infections, medications and foreign antigens like tattoo. CLH mimics lymphoma clinically and histologically. Progression to cutaneous lymphoma is described in the literature, and is hypothesized to be due to persistent antigenic stimulation (1).

We present an interesting case of idiopathic CLH that failed to respond to multiple medical and surgical interventions. Ultimately, the patient demonstrated a significant therapeutic response to treatment with dupilumab. This case highlights the potential role of targeted biologics in the treatment of idiopathic CLH.

Materials & Methods:

We present the case of a 32-year-old gentleman, first presenting in 2011. The patient developed a persistent, thickened and painless lesion on the right cheek with level II cervical lymphadenopathy. Facial lesion was soft, non-scaly and erythematous on examination. Differential diagnosis included infections (e.g., Borrelia), Kimura disease, lupus panniculitis/tumidus and cutaneous lymphoma. In 2013, patient's history was complicated by secondary eczema on the body.

Histopathological and Immunohistochemistry studies revealed reactive lymphoid hyperplasia/B cell pseudolymphoma on multiple biopsies. In 2014, the presence of germinal centre hyperplasia, peripheral eosinophilia raised the possibility of Kimura disease. However, further histological review advised against this diagnosis due to the absence of key histopathological features of Kimura disease, such as eosinophilic infiltrate and microabscesses formation.

A single focus of reactive Langerhans cell histiocytosis showed up in a review of the case in 2023. This led to query the diagnosis of systemic Langerhans cell histiocytosis, however further hormonal tests and PET-CT ruled out multisystem involvement. Patient was extensively investigated using viral and bacterial serology, viral PCR and CT-NCAP, all were negative for malignancy or chronic infections.

Results:

Multiple medical and surgical treatments were attempted over a 10-year period which were ineffective with the exception of Oral Corticosteroids at doses > 20mg per day. These included methotrexate, intralesional steroid, interferon-alpha, cyclosporine, Fumaderm, Intralesional and Intravenous Rituximab. Radiotherapy for local control was trialled following a combined dermatology and oncology MDT. Multiple specialties, including Dermatology, Haematology, Infectious Disease, Oncology and Plastic Surgery, were involved throughout disease course.

In 2024, initiation of dupilumab (IL-4Rα blocking monoclonal antibody) due to severe eczema led to significant and consistent improvement both in eczema and facial lesion. Dupilumab inhibits B-cell proliferation by blocking IL-4 and IL-13, leading to reduced STAT6 activation and increased B-cell apoptosis (2).

Conclusion:

This case demonstrates how challenging the diagnosis and treatment of idiopathic CLH can be. Dupilumab cleared effectively both eczema and CLH lesions. This case highlights a future potential role of targeted biologics and the need to explore immunomodulatory strategies in similar cases.

CLH: Cutaneous Lymphoid hyperplasia