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Case Report

Jessner's Lymphocytic Infiltration of the Skin (JLIS): A Rare Forehead Skin Involvement

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ABSTRACT

Rare cutaneous condition of Jessner's lymphocytic infiltration of the skin (JLIS) is a chronic, benign T-cell infiltrative disorder. It is a skin disease manifesting as red-colored papules or plaques on the face, neck and back. This lesion can be single or multiple with frequent recurrence. We report the case of a 35-year-old male, who presented with on and off multiple, well defined, discrete, erythematous plaques over forehead skin with 15 years duration, exacerbated during winters. The gold standard, 'skin biopsy' confirmed the diagnosis of JLIS over the forehead.

Keywords: Jessner's lymphocytic infiltration of the skin, Forehead

INTRODUCTION

Jessner lymphocytic infiltration of the skin (JLIS) is a rare condition with unknown incidence and prevalence in both the U.S. and worldwide. The disease onset is usually between 30 and 50 years of age; however, there are cases reported in children, though they are less common.[1]

Jessner's lymphocytic infiltration of the skin is the term used to describe a rare, benign cutaneous condition characterized by papular or plaque-like eruptions that commonly involve face, neck and trunk in sun-exposed areas.[1] The other names of Jessner's lymphocytic infiltrate are benign lymphocytic infiltrate, Jessner's kanof lymphocytic infiltrate, and benign chronic T-cell infiltrative disease. Jessner lymphocytic infiltration of the skin (JLIS) remains unknown. There may be a genetic/hereditary component due to multiple reported familial cases of the disease. Another possible etiology is autoimmune. The plasmacytoid dendritic cells present in JLIS are known to play a role in systemic lupus erythematosus.[2] JLIS represents a characteristic CD8+ polyclonal reactive skin condition in immunohistochemistry.

CASE HISTORY

A 35-year-old Indian army man presented with the chief complaints of red-colored lesions over forehead for 15 years. Patient reported that such lesions appear during cold weather of the winters and wane off later. On cutaneous examination multiple, well defined, discrete, erythematous plaques over forehead were seen (Figure 1a&b). The complete blood count was under normal range. Renal function tests, liver function tests, thyroid function tests, erythrocyte sedimentation rate, antinuclear antibody test; anti-Ro and anti-La antibodies were also normal.

Then lastly skin biopsy was done, and biopsy specimen was sent for histopathology examination. On gross examination, the received skin covered tissue bits, totally measured 0.3x0.2x0.2 cm in size. All tissue processed in one block and subjected to formalin fixed paraffin embedded sectioning (FFPE).

On Microscopic examination, H&E sections showed orthokeratosis, mild hyperkeratosis, hypergranulosis and mild acanthosis. Upper dermis showed perivascular and peri-adnexal mild mononuclear infiltrate. Adnexal tissue structures noted. Marked lymphoid collection noted in upper dermis at places. Plasmacytoid monocytes were seen within the lymphoid collections. There was no sign of granuloma formation or histiocytes containing Leishman bodies in sections studied. Subcutis was unremarkable.

Topical corticosteroids with oral hydroxychloroquine were found to be effective for the treatment of JLIS in our case on follow-up.

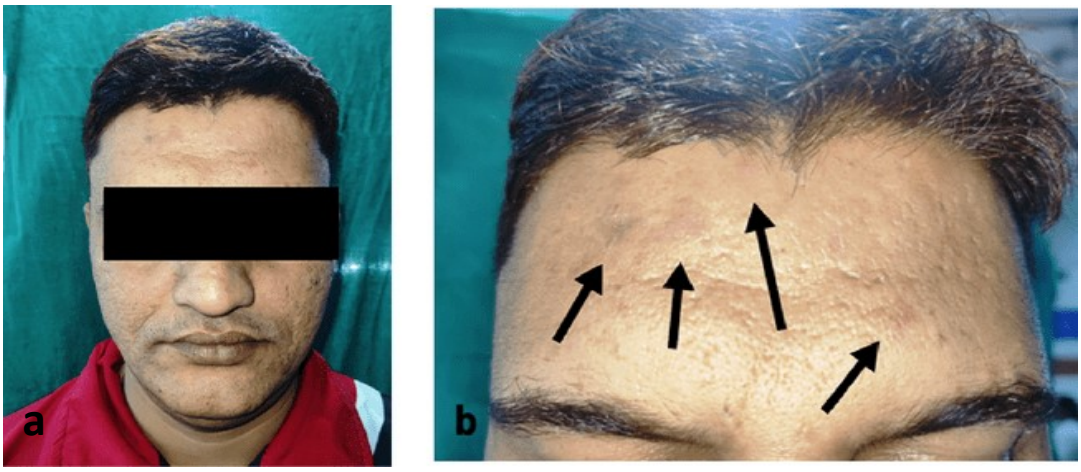

Figure 1: (a) Clinical photograph showing entire face **(b)** Clinical photograph showing multiple, well defined, discrete, erythematous plaques (*arrows*) over forehead skin

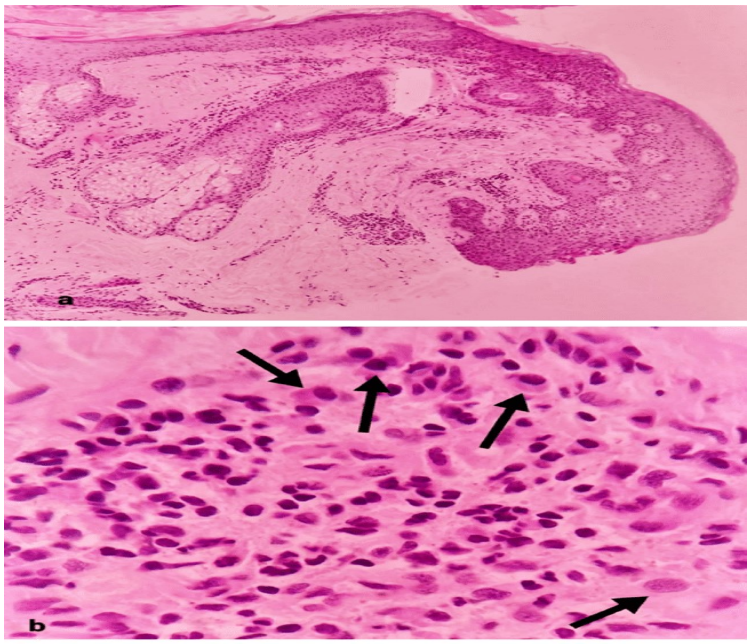

Figure 2: (a) Microphotograph shows (H and E, X40) orthokeratosis, mild hyperkeratosis, hypergranulosis and mild acanthosis without lymphocytic exocytosis. Upper dermis shows mild mononuclear infiltrates located peri-vascularly and periadnexally. Marked lymphoid collection noted in upper dermis at places (H&E, X40) **(b)** Plasmacytoid monocytes *(arrows)* noted within lymphoid collections (H&E, X400)

DISCUSSION

Jessner's lymphocytic infiltrate of the skin (JLIS) described by Jessner and Kanof in 1953 is known as a benign chronic T-cell disorder. It mostly affects middle aged adults with few reported cases.[3] JLIS classically presents as red tumid nodule/ plaque over face, neck, upper trunk and back with slight seasonal variation. It gets precipitated during winters, as in this case.

The lesions are smooth, raised non-scaling erythematous nodules or plaques. Some other diseases having similar patchy dermal infiltration are discoid lupus erythematous, leprosy, polymorphous light eruption, mycosis fungoides, granuloma annulare, lymphocytoma cutis and lymphoma must be ruled out to reach a final diagnosis.

JLIS occurs in sun exposed area whereas discoid lupus erythematosus (DLE) may or may not occur in sun exposed area. Hyperkeratosis is mostly absent in JLIS whereas it is present in DLE.[4] However, our case had hyperkeratosis owing to the waxing and wanning episodes, skin scratch and chronicity of lesions over 15 years.

Direct immune-fluorescence (DIF) plays a significant role in making a final diagnosis which is positive in Discoid lupus erythematosus and negative in JLIS. Other differentials include mycosis fungoides, which is one of the common types of cutaneous T-cell lymphoma also called as a mature T-cell non-hodgkin's lymphoma caused by proliferations of small to medium sized lymphocytes with cerebriform nuclei.

Histopathologically JLIS is characterized by a superficial and deep, primarily perivascular, sleeve-like lymphocytic dermal infiltrates with predominance of small mature polygonal lymphocytes and without epidermal involvement. Deposits of mucin in the reticular dermis have been described, however not seen in our case.[5]

A variety of treatments has been tried, with variable and often limited success, including topical, intra-lesional or systemic corticosteroids, antimalarial, thalidomide, tetracyclines, cryotherapy and photo protections. A few cases of successful treatment with dapsone, auranofin and chemotherapy have been reported. Treatment with hydroxychloroquine is found effective for the treatment of JLIS.

One case of JLIS disease has been treated by pulsed dye laser. Pulsed dye laser has been used in cutaneous lupus and annular granuloma. Selective photo thermolysis allows photocoagulation of dilated vessels. Pulsed dye laser at 595nm could offer a valuable therapeutic alternative, and even a first line treatment with no evident side effects.[6]

Lymphocytic infiltration of the skin and reticular erythematous mucinosis is characterized histologically by an inflammatory cutaneous lymphocytic infiltrate similar to the histological appearance of pseudo-lymphoma. Lymphocytic infiltration of skin and reticular erythematous mucinosis seems to represent clinico-pathological reaction patterns. Most cases of both conditions constitute hidden variants of lupus erythematosus. Lymphocytic infiltration of skin in contrast to reticular mucinosis frequently comprises pseudolymphomatous reaction including borrelial lymphocytoma.

JLIS clinically manifests in face, in particular on the cheeks and earlobes, but also on the neck, upper trunk, or the proximal extremities of middle-aged adults. Our case had forehead skin involvement. Lesions consist of solitary or multiple erythematous, discoid plaques that are often arranged in crescent or ring formations. They persist for months or years and resolve spontaneously without treatment, but often recur.[7]

JLIS skin lesion on H&E sectioned biopsy reveals well demarcated dense perivascular infiltrate and peri adnexal infiltrate in the dermis composed of mature lymphocytes and these lymphocytes showed immunoreactivity to CD4 antibodies with immunohistochemistry staining.[6,7] IHC was not done in our case, due to non-availability and cost-restraint. There is no sign of skin granuloma formation or histiocytes containing Leishman bodies or other signs of Leishmaniasis in JLIS.[8]

JLIS is a clinically and histologically distinct disease entity. Conflicting results have been reported concerning its differentiation from cutaneous lupus

erythematosus and polymorphous light eruption, its relationship to palpable migratory arciform erythema and its classification as a CD4+ T-cell lymphoproliferative disease.[9]

This disease is diagnosed by biopsy revealing perivascular and peri adnexal clusters of plasmacytoid monocytes within the dermis (just like in our case), sometimes extending into the subcutaneous tissue. The subcutaneous plasmacytoid monocytic infiltrate was not seen in our case. The plasmacytoid monocyte cells, also known as plasmacytoid dendritic cells, play a vital role in the induction of autoimmune diseases and other skin diseases. While their presence, along with an inappropriate CD8+ T-cell predominant lymphoid infiltrate, provides a definitive diagnosis of JLIS, the cause of this disorder remains unknown. There is difference between JLIS and lupus erythematosus tumidus. In JLIS, there is more frequent involvement of back skin and in lupus erythematosus tumidus there is more frequent face involvement with plaques showing non-caseating skin granulomas.[10]

CONCLUSIONS

JLIS is a rare, cyclic, chronic skin disorder associated with waxing and waning episodes triggered during winters and possibly genetic disposition. The prognosis is good on treatment with local corticosteroids and oral hydroxychloroquine.

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