Case Report

The Puzzle of Papules Over Face and Extrafacial Areas: A Rare Case of Disseminated Idiopathic T-Cell Pseudolymphoma

Abstract

Papular lesions over face have a wide range of differential diagnoses. The clinical differentiation among these is difficult and often requires clinicopathological correlation. We hereby report the case of a 28 year old female presenting with numerous skin colored papular lesions over face, pinna and upper limbs.

Keywords: *Cutaneous lymphoid hyperplasia, lichenoid pseudovesicular papular eruption on nose, lymphocytoma cutis, lymphocytic infiltrate, papules over nose*

Sumit A. Hajare, Vaishali H. Wankhade[1], Gitesh U. Sawatkar[2], Rajesh Pratap Singh[1]

Department of Dermatology, Indira Gandhi Government Medical College, [1]Department of Dermato-Venereo-Leprology, Government Medical College, [2]Department of Dermatology, All India Institute of Medical Sciences, Nagpur, Maharashtra, India

Introduction

Papular lesions over face have varied differential diagnoses including granulomatous rosacea, acne vulgaris, perioral dermatitis, Jessner's lymphocytic infiltrate of the skin (JLIS), papular sarcoidosis, polymorphic light eruption, lupus erythematosus, adenoma sebaceum, pseudolymphoma, among others. The clinical differentiation between these is difficult and often requires clinicopathological correlation. We hereby report a case presenting with multiple skin colored papules over face, pinna, and upper limbs.

Case History

A 28-year-old housewife presented with multiple asymptomatic papules over face and upper limbs since six years. The lesions developed initially over nose and gradually progressed to involve the cheeks, forehead, pinna and upper limbs since one year. The patient complained of mild burning sensation on sun exposure and exacerbation of lesions in summer. The patient denied history of any systemic illness, seizure disorder, history of similar lesions in family, any drug intake during or prior to development of lesions.

Cutaneous examination revealed multiple 2- to 5-mm-sized skin colored to erythematous, translucent, firm papules present bilaterally symmetrical over nose, cheeks, and forehead extending into frontal hairline with sparing of periorbital area [Figure 1]. Similar lesions were present bilaterally over pinna, pre- and post-auricular areas [Figure 2a and b]. The patient had numerous erythematous papules of size 2–5 mm interspersed with few lichenoid papules over both forearms and dorsum of hands [Figure 2c and d]. Examination of other cutaneous sites, mucosae and lymph nodes were normal. Systemic examination was normal. On the basis of clinical examination, the differential diagnoses of sarcoidosis, Jessner's lymphocytic infiltrate of the skin, pseudolymphoma, rosacea with extrafacial involvement were considered. The routine blood investigations and chest radiograph were normal. Anti-nuclear antibody and Mantoux tests were negative. Levels of calcium and serum angiotensin converting enzyme were normal. Serological tests for T. pallidum, Human Immunodeficiency Virus (HIV) and Borrelia burgdorferi were negative.

Skin biopsy was performed from lesions over forehead and pre-auricular area which showed a normal epidermis with clear subepidermal zone, and dense and diffuse lymphocytic infiltrate throughout the dermis. The subcutaneous layer was normal [Figure 3a and b]. There was no evidence of granuloma

How to cite this article: Hajare SA, Wankhade VH, Sawatkar GU, Singh RP. The puzzle of papules over face and extrafacial areas: A rare case of disseminated idiopathic T-cell pseudolymphoma. Indian Dermatol Online J 2021;12:312-5.

Received: 19-Jul-2020. **Revised:** 07-Aug-2020. **Accepted:** 05-Sep-2020. **Published:** 02-Mar-2021.

Address for correspondence:
Dr. Vaishali H. Wankhade, Department of Dermato-Venereo-Leprology, Government Medical College, Nagpur - 440 003, Maharashtra, India.
E-mail: vshingade73@yahoo.in

Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ_568_20

Quick Response Code:

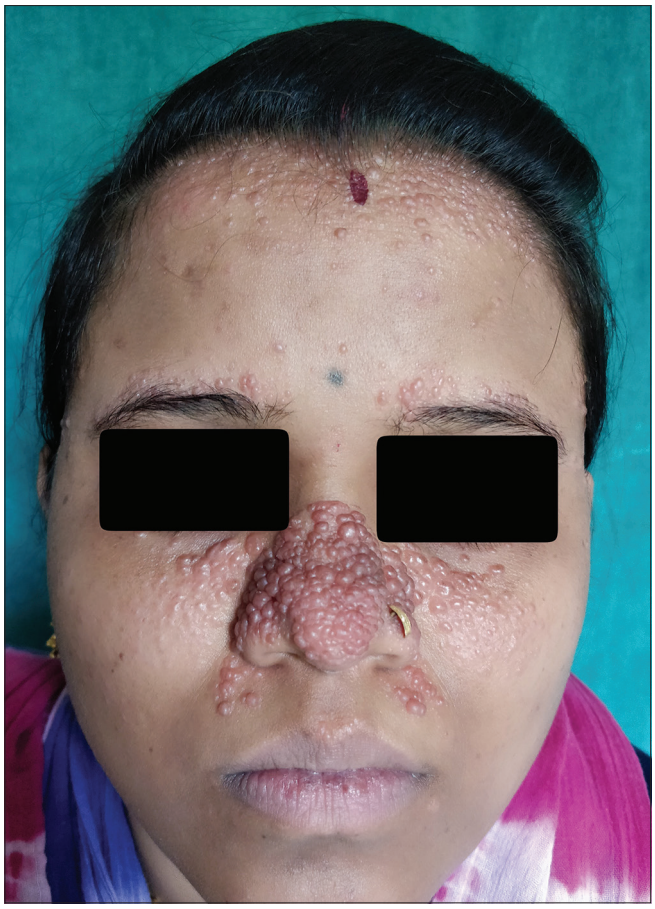

Figure 1: Multiple skin colored to erythematous, translucent, firm papules were present bilaterally symmetrical over nose, cheeks, and forehead

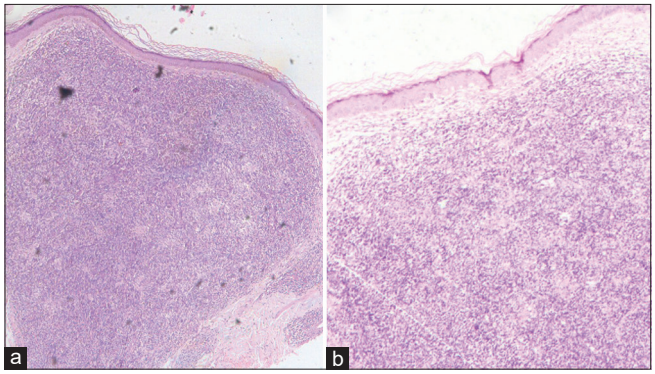

Figure 3: Histopathological image showing (a) Normal epidermis with clear subepidermal zone, and dense and diffuse lymphocytic infiltrate throughout the dermis (H and E, ×20). (b) Similar findings seen on higher magnification (×40)

formation or atypical lymphocytes and reticulin staining was negative. On clinicopathological correlation, provisional diagnosis of pseudolymphoma was made and immunohistochemistry (IHC) studies were advised. On IHC staining, the infiltrate was strongly positive for CD8 and CD3 cells (pan T-cell marker), weakly positive for CD5 and CD7 cells and negative for CD4 T-cells [Figure 4a-d]. Along with predominant involvement by T-cells, infiltrate

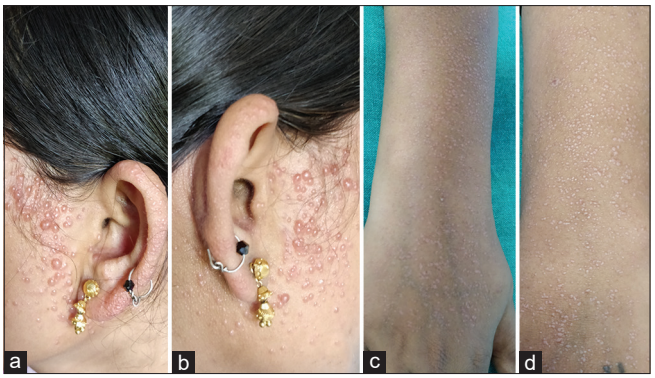

Figure 2: (a and b) Clinical image showing skin-colored papules present bilaterally over pinna, pre- and post-auricular areas. (c and d) Multiple erythematous and lichenoid papules over both forearms and dorsum of hands

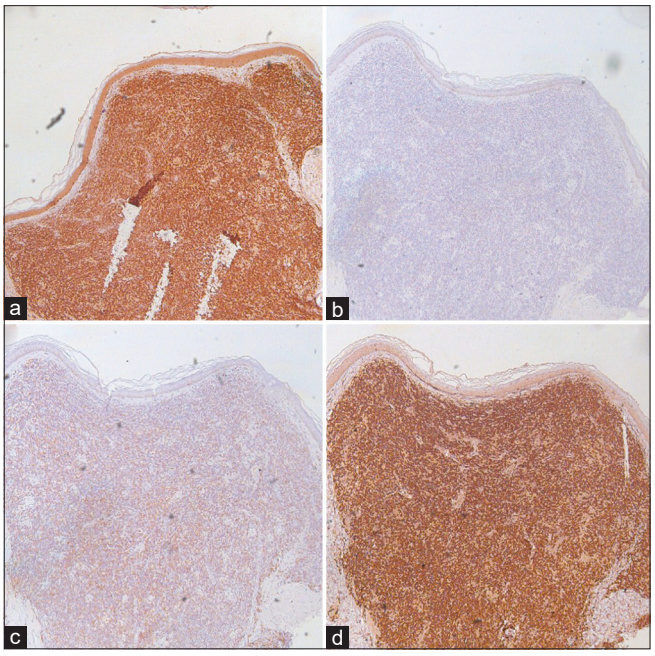

Figure 4: Immunohistochemistry staining for (a) CD3+ T cells, (b) CD4+ T cells, (c) CD5+ T cells, (d) CD8+ T cells showing infiltrate strongly positive for CD3+ and CD8+ T cells

was also weakly positive for CD20 (pan B-cell marker) and CD68 (histiocytes) cells. Ki-67 was weakly positive and showed normal degree of proliferation and bcl-2 was negative [Figure 5a-d]. On the basis of clinical, histopathological and IHC findings, the final diagnosis of disseminated idiopathic T-cell pseudolymphoma was confirmed.

The patient was advised avoidance of cosmetics along with strict photoprotection and use of topical sunscreen gel. She was started on treatment with oral minocycline 100 mg once daily. The lesions showed improvement after three months of treatment and majority of lesions over face and limbs resolved except a few lesions over nose. However, the patient stopped treatment after three months and was lost to follow up for a period of six months after which

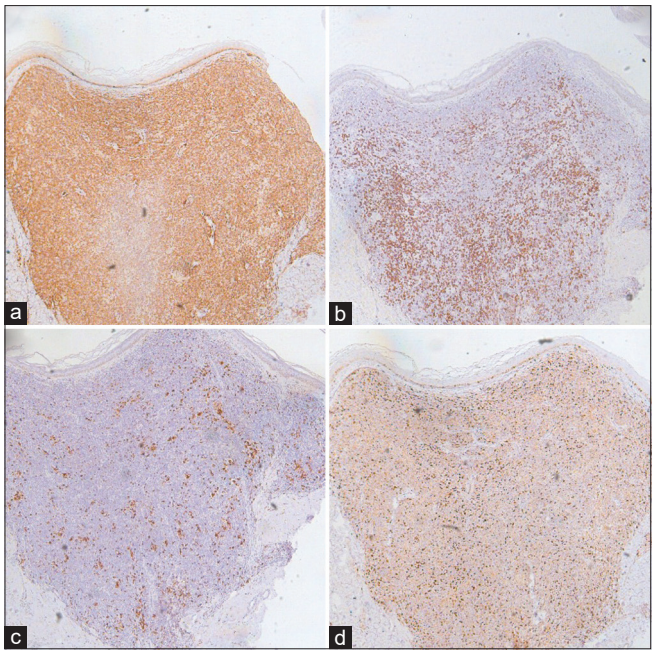

Figure 5: Immunohistochemistry staining for (a) bcl-2, (b) CD20+ B cells, (c) CD68+ histiocytes, (d) Ki-67 showing infiltrate weakly positive for CD20+ B cells and CD68+ histiocytes

she presented with recurrence of lesions. The patient was treated with oral prednisolone 40 mg once daily, which was tapered over a period of three months along with oral hydroxychloroquine 300 mg once daily and topical sunscreen continued for six months. There was an excellent improvement after six months of treatment [Figures 6 and 7a-d] with complete resolution of majority of lesions. Few lesions over face healed with pinpoint depressed scars. The papules over nose which were initially recalcitrant to treatment, flattened after six months. The patient was counseled regarding regular treatment and repeat biopsy and IHC at regular intervals to rule out lymphoma.

Discussion

Cutaneous pseudolymphoma (PSL) is a heterogeneous group of disorders with lymphocyte-rich infiltrate and simulates cutaneous lymphomas clinically and/or histologically. Cutaneous PSL was first termed as Sarcomatosis cutis by Kaposi (1890). The term cutaneous pseudolymphoma was later coined by Burg. PSLs have also been described as lymphocytoma cutis, cutaneous lymphoid hyperplasia, cutaneous prelymphomas, and lymphadenosis benigna cutis.[1]

The cellular components of skin associated lymphoid tissue (SALT) in the dermis include CD4+ T-cells, CD8+ T-cells, dendritic cells and rarely macrophages and B cells.[1] Persistent antigenic stimulation of SALT results in formation of reactive lymphoid infiltrates known as cutaneous PSLs which are benign in nature. Chronic inflammation and oxidative damage may result in acquired

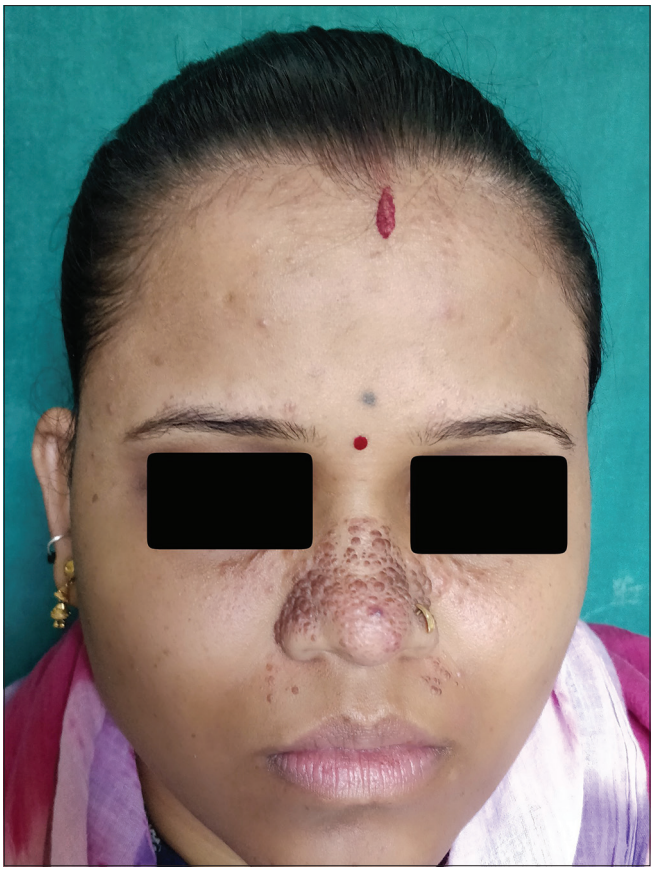

Figure 6: Post treatment clinical image of face showing good response to treatment except few lesions over nose

genetic changes leading to formation of atypical lymphoid proliferations and lymphomas. PSLs occur as a result of recruitment and selective accumulation of lymphocytes and are classified as either B-cell, T-cell or mixed as per the predominantly stimulated arm of immune response.[1]

PSL is idiopathic in majority of the cases. However, association with exposure to foreign antigens such as arthropods (bites, stings, infestations), infections (Herpes viruses, Borrelia burgdorferi, Treponema pallidum, Molluscipoxvirus and HIV), tattoos, acupuncture, trauma, gold piercing, injected vaccinations, hyposensitization injections, photosensitivity, and medications have been described.[2,3] In the present case, history of photosensitivity and gold piercing was elicited. The localization of the lesions over contours of face and extensors of upper limbs suggests a possible role of sun exposure as a trigger or exacerbating factor.[4]

Cutaneous PSL often presents as a solitary nodule involving the exposed areas such as head and neck region and upper extremities.[1,2,4] Multiple lesions can be present, either aggregated in clusters (agminated form) or as disseminated papules (miliarial form). Disseminated PSL is very rare with very few cases reported in literature. Moulonguet *et al.* described the miliarial-type of pseudolymphoma, in two women who had transluscent micropapules over

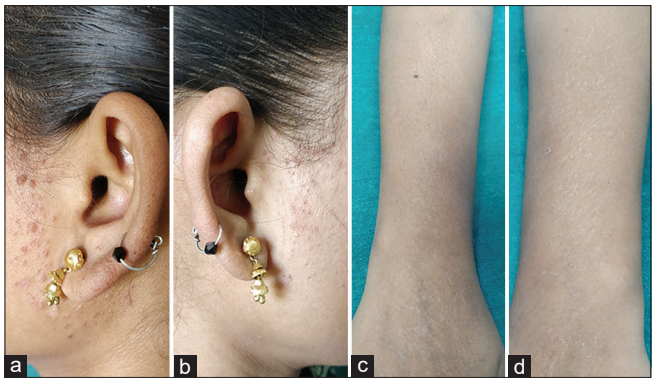

Figure 7: Post treatment clinical images of (a and b) bilateral pinna, pre- and post-auricular areas, and (c and d) both upper limbs showing resolution of lesions

face, sides of neck and forearms. These cases showed lymphoid follicle formation on histopathology with predominance of B-cells and presence of few CD3+ T-cells suggestive of B-cell PSL.[4,5] Jain *et al.* reported a case of T-cell predominant cutaneous lymphoid hyperplasia in a 24-year-old female who had skin-colored papules over nose, malar area and forehead. Histopathology of the patient showed focal epidermal atrophy, a subepidermal grenz zone and nodular lymphocytic infiltration in upper and mid dermis and CD3+ T-cell predominance on IHC.[6] In present case, histopathology showed dense and diffuse lymphocytic infiltrate in dermis and predominance of CD8+ and CD3+ T-cells, and few CD20+ B-cells and histiocytes suggestive of T-cell PSL. Singh S. *et al.* recently described cases with similar morphological features as lichenoid pseudovesicular papular eruption on nose.[7] These cases had similar clinical presentation along with focal nodular lymphoid aggregates in the papillary dermis, epidermal basal cell degeneration and focal epidermal atrophy. However, complete immunohistochemistry profile was not done in these cases and showed equal presence of CD3 and CD20 cells as compared to predominant CD8 and CD3 cells in the present case. Further study and evaluation of the histopathological characteristics and IHC staining in such cases will be helpful in understanding the disease and correct classification.

Histologically, T-cell PSLs may present as band-like pattern (mycosis fungoides-like), diffuse pattern (mimicking tumor stage of mycosis fungoides), nodular pattern, polypoid angiomatoid pattern, and pseudolymphomatous folliculitis-like pattern.[1,2] In T-cell PSLs, there is lack of considerable B-cell component and CD4+ T helper-inducer cells are usually sparse.[1] In the present case, CD8+ T cells were predominant with absence of CD4+ T cells.

Dense and diffuse T-cell infiltrate occupying the near-totality of the dermis can be seen in drug-induced and idiopathic-T-cell pseudolymphoma and clonal lymphoproliferative disorders.[8] Sarcoidosis, JLIS, lupus miliaris disseminatus faciei and steroid acne can be excluded on the basis of histological profile.[4] Rosacea with extrafacial involvement may have a similar clinical presentation but can be differentiated by presence of inflammatory papules and poorly organized epithelial granulomas and follicular pustules in the biopsy.[9] Miliarial form of primary cutaneous follicle center lymphoma represents the main clinical and histopathological differential diagnosis but can be differentiated on immunohistochemistry.

The clinical course of cutaneous PSL is variable. Some cases may show regression spontaneously, but many persist for several months or years. Recurrences may occur after re-exposure to the inducing agent. Solitary lesions can be treated by surgical excision, cryotherapy, topical tacrolimus and topical or intralesional corticosteroid. For disseminated lesions, systemic corticosteroids, intralesional or systemic interferon alpha, oral hydroxychloroquine, and minocycline can be used.[2,3]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Hussein MR. Cutaneous pseudolymphomas: Inflammatory reactive proliferations. Expert Rev Hematol 2013;6:713-33.
2. Mitteldorf C, Kempf W. Cutaneous Pseudolymphoma. Surg Pathol Clin 2017;10.455-76.
3. Engin B, Songür A, Kutlubay Z, Serdaroğlu S. Lymphocytic infiltrations of face. Clin Dermatol 2014;32:101-8.
4. Moulonguet I, Ghnassia M, Molina T, Fraitag S. Miliarial-type perifollicular B-cell pseudolymphoma (lymphocytoma cutis): A misleading eruption in two women. J Cutan Pathol 2012;39:1016-21.
5. Thomas EA, Masih K, Sadiq S. Disseminated lymphadenosis benigna cutis. Indian J Dermatol 2006;51:62-3.
6. Jain A, Majumdar B, Sen D, Sen S, Mishra P, Samanta A. Asymptomatic papules over central and pericentral areas of the face. Indian Dermatol Online J 2015;6:198-200.
7. Singh S, Singh A, Mallick S, Arava S, Ramam M. Lichenoid pseudovesicular papular eruption on nose: A papular facial dermatosis probably related to actinic lichen nitidus or micropapular polymorphous light eruption. Indian J Dermatol Venereol Leprol 2019;85:597-604.
8. Charli-Joseph YV, Gatica-Torres M, Pincus LB. Approach to cutaneous lymphoid infiltrates: When to consider lymphoma? Indian J Dermatol 2016;61:351-74.
9. Pereira TM, Vieira AP, Sousa Basto A. Rosacea with extensive extrafacial lesions. Int J Dermatol 2008;47:52-5.