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Co-Occurrence of Lichen Planus and Alopecia Areata: A Possible Role of Plasmacytoid Dendritic Cells

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ABSTRACT

Plasmacytoid Dendritic Cells (pDCs), also called Natural Interferon (IFN)-producing cells, represent a specialized cell type within the innate immune system that are normally found in lymphoid tissues and possess both antigen-presenting and immune markers. These cells release high amounts of type I interferon (IFN-I) providing, protective immunity at the level of the skin by regulated sensing of microbial- or self-nucleic acids. Upon skin damage, excessive sensing by pDCs may elicit IFN-driven inflammatory/autoimmune skin disorders. IFN-I production by plasmacytoid dendritic cells plays an immunogenic role in autoimmune skin disorders. We present a case report of the coexistence of two common autoimmune skin conditions, lichen planus (LP) and alopecia areata (AA), and hypothesize a possible role of shared plasmacytoid dendritic cells - interferon (pDC-IFN) pathway, in various autoimmune skin disorders.

Keywords: Alopecia areata, Lichen planus, Coexistence, Plasmacytoid Dendritic Cells, Interferons

INTRODUCTION

A 14-year-old female presented with multiple dark-colored, itchy lesions on her legs for 5 months. On examination, multiple violaceous papules to plaques of varying sizes (1 cm-5 cm) were present on the extensor aspects of the legs, forearm, and dorsum of feet bilaterally (Figure 1-3). Skin biopsy from the lesion showed hyperkeratosis, hypergranulosis, vacuolar degeneration of the basal layer, band of dense lymphocytic inflammatory infiltrate in the

papillary dermis, with perivascular histiocytic infiltrate confirming the diagnosis of Lichen Planus (LP) (Figure 4 and 5). She was started on topical corticosteroids, antihistamines and emollients. After 3 months the patient had aggravation of LP with patchy hair loss over the scalp. On examination multiple, smooth alopecic patches of varying sizes, the largest being 4 x 3 cm, were noticed on the scalp. She was diagnosed clinically with Alopecia Areata (AA) (Figure 6). Investigations like complete blood count, liver function test, thyroid profile, anti-nuclear antibody, rheumatoid arthritis factor, C-reactive protein, ESR, VDRL, HBV, HCV, and urine microscopy were normal. Given progressing lesions of LP and AA, she was started on oral mini pulse therapy-betamethasone 5 mg twice weekly and was advised for follow-up. Good response was noticed by four weeks with resolution of LP and regrowth of hair over few patches.



Figure 1 Multiple violaceous papules to plaques of varying sizes over the extensor aspects of both legs bilaterally



Figure 2 Multiple violaceous papules to plaques over the dorsum of feet bilaterally



Figure 3 Few erythematous papules over the wrist area

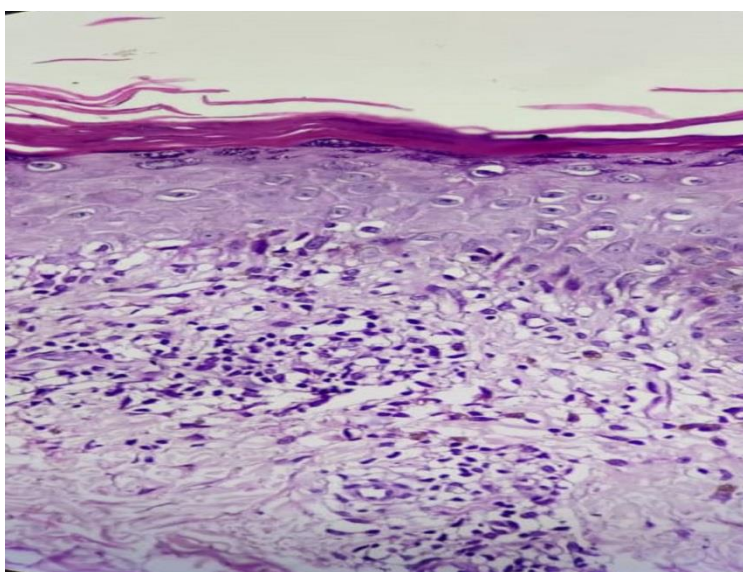


Figure 4 Photomicrograph showing hyperkeratosis, parakeratosis, hypergranulosis, band of dense lymphocytic inflammatory infiltrate, histiocytes admixed with congested blood vessels, along with periadnexal inflammatory infiltrate (H and E, x 40)

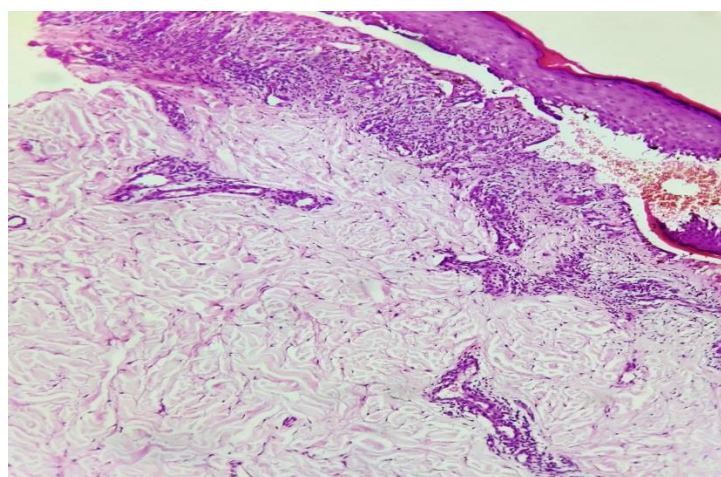


Figure 5 Photomicrograph showing hyperkeratosis, parakeratosis, hypergranulosis, artifactual cleft formation between the epidermis and papillary dermis (Max Joseph space), band of dense lymphocytic inflammatory infiltrate, histiocytes admixed with congested blood vessels, along with periadnexal inflammatory infiltrate (H and E, x 10)



Figure 6 Multiple well-demarcated, smooth, bald, round alopecic patches of hair loss of varying sizes on the scalp

LP and AA are autoimmune dermatoses and are associated with other conditions like diabetes mellitus, vitiligo, autoimmune thyroid diseases, etc to name a few. So far, only 3 reports of co-localization of AA and LP have been published in the literature [1-3]. Till now, no case report of the coexistence of LP and AA has been reported. Plasmacytoid Dendritic Cells (pDCs) are specialized dendritic cells exhibiting plasma cell morphology, expressing CD4, CD123, HLA- DR, Blood-Derived Dendritic Cell Antigen-2 (BDCA-2), Toll-Like Receptor (TLR)7 and TLR9 within endosomal compartments and their role in autoimmune diseases is gaining traction recently. On activation, they produce type I IFN, against pathogenic agents and link the innate and adaptive immunity by controlling the function of myeloid dendritic cells, T, B, and natural killer cells. pDCs are absent in normal skin but infiltrate when injured, thereby contributing to the pathogenesis of inflammatory dermatoses (like LP and AA). Vries et al found pDCs in close approximation to the basal layer in lesional LP, corresponding to lymphocytic infiltration [4]. It has been hypothesized that some common antigenic determinant may be a triggering factor for the onset of both diseases, and thus, a primary autoimmune process directed against basal epidermal cells in LP could have possibly resulted in disruption of hair follicle immune privilege zone thereby exposing the hidden antigens from hair follicles, leading to pDCs recruitment, production of IFN-gamma and resulting in aggravation of LP and occurrence of secondary autoimmune response i.e., AA [5]. This explains the coexistence of LP and AA in our case report. This rare case of sequential occurrence of LP followed by the AA has not been reported previously and might offer possible theories which contribute to the literature on T cell-mediated autoimmune disorders.

DECLARATIONS

Conflict of Interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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