Journal of Pakistan Association of Dermatologists 2012;**22**:55-58.

Case Report

Association of alopecia areata with segmental vitiligo and segmental lichen planus: a case report

Piyush Kumar*, Anubhav Gosai, Ashim Kumar Mondal**, Panchami Debbarman**, Niharika Ranjan Lal**, Ramesh Chandra Gharami***, Arunasis Maiti†**

*Dermatology Department, Katihar Medical College, Katihar. India
**Dermatology Department, Medical College, Kolkata. India
†Dermatology Department, Midnapore Medical College & Hospital, Midnapore, India

Abstract Alopecia areata (AA) is a chronic inflammatory condition that affects the hair follicles and sometimes, the nails. It is believed that AA is caused by T cell-mediated autoimmune response and increased frequency of other autoimmune diseases in persons suffering from AA supports this hypothesis. A 12-year-old boy presented with extensive alopecia areata with pitting in all nails for 6 years. He had developed lesions of vitiligo and lichen planus on trunk in a segmental manner. While the association of alopecia areata and vitiligo as well as lichen planus is well known, segmental nature of both vitiligo and lichen planus is quite rare.

Key words
Alopecia areata, segmental vitiligo, segmental lichen planus.

Introduction

Alopecia areata (AA) has been reported in association with many autoimmune conditions and list of such conditions is increasing with each passing day. Such an association carries prognostic importance too, as presence of other autoimmune conditions is considered poor prognostic factors.[1,2] Our case, a 12-year-old boy with AA, developed segmental vitiligo and segmental lichen planus (LP). Though association with vitiligo and LP are well known,[1] segmental nature of these conditions is quite rare. We report our case for its rare association with segmental vitiligo and segmental lichen planus.

Case report

A 12-year-old boy presented with complaints of asymptomatic loss of scalp hair for 6 years and hyperpigmented scaly lesions on trunk for 1 year. The lesion on scalp started as a small area of hair loss and progressed to present status. There was history of complete recovery; however, new areas of hair loss used to appear. There was no history of any lesions on scalp prior to development of hair loss. He developed some hyperpigmented scaly lesions on trunk one year back. These lesions were slightly itchy. He had received treatment from a local physician with no improvement. Some 2 months back, he developed hypopigmented lesions on trunk and upper arm. They were asymptomatic and were increasing in number; however, they were limited to particular part of trunk only. Rest of the history was non-contributory and no other family member was having similar skin lesions.

On examination, well-defined areas of alopecia on scalp were noted and it was non-scarring in nature. Ophiasis pattern was well appreciable (**Figure 1**). On trunk, two types of lesion were seen. Some of the lesions were

Address for correspondence
Dr. Piyush Kumar,
Assistant Professor
Dermatology Department, Katihar Medical College, Katihar. India
Ph# +91 8987280259
Email: docpiyush@gmail.com

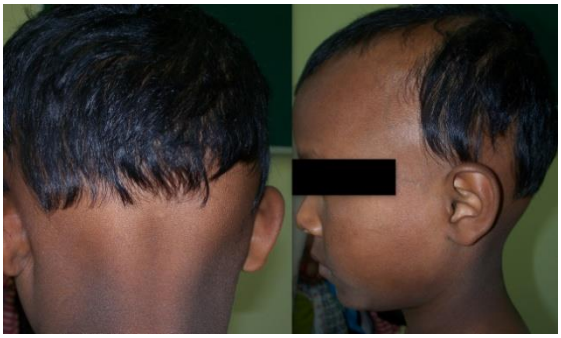

Figure 1 Alopecia areata. Note Ophiasis pattern.

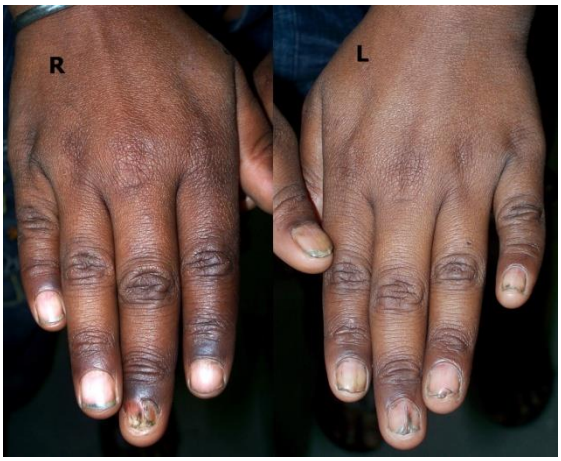

Figure 3 Fine pitting in all nails. Note pterygium of right middle finger and longitudinal splitting of left middle finger.

hypopigmented macules and patches without any erythema and scaling. The central parts of some of these lesions were completely depigmented. These lesions were present in a segmental manner over left upper back and extending on left arm. The other type of lesion was hyperpigmented scaly papules and plaques present on left upper trunk in a segmental manner. Similar hyperpigmented plaques were present on left upper chest too (**Figure 2**). No other lesions were present elsewhere on the body. All nails showed fine pitting. The nail of right middle finger showed dorsal pterygium, while that of left middle finger showed longitudinal splitting (**Figure 3**). There was no pallor and thyroid gland appeared to be normal in size. Rest of the mucocutaneous and systemic examination was unremarkable. Complete hemogram, blood sugar, and thyroid profile did not reveal any abnormality. KOH mount of the scrapings from the hypopigmented lesions did not reveal

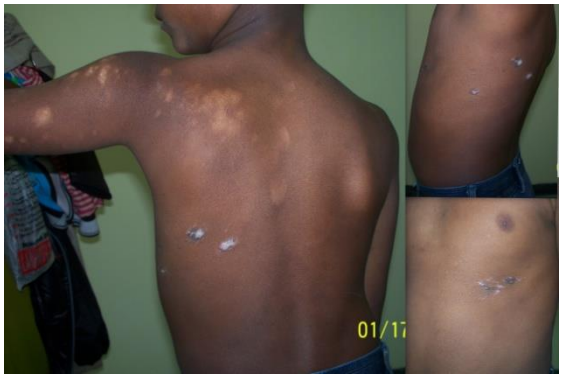

Figure 2 Lesions of vitiligo and lichen planus on trunk in a segmental manner.

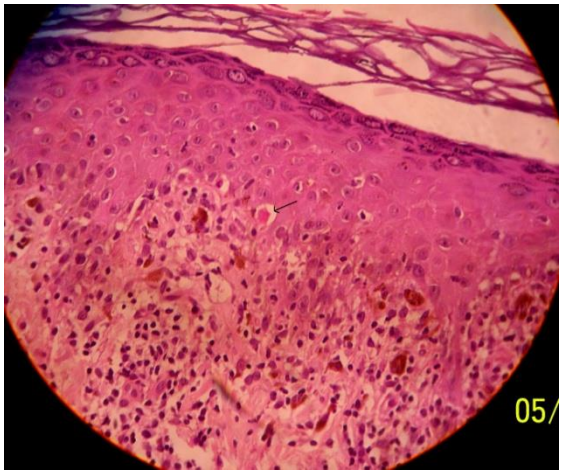

Figure 4 Photomicrograph showing hyperkeratosis, focal hypergranulosis, band like infiltration with mononuclear cells in upper dermis and basal layer vacuolar degeneration. Note colloid body (arrow).

any fungal elements. Histopathology from the hyperpigmented lesions showed hyperkeratosis, focal hypergranulosis, acanthosis, dense lichenoid mononuclear cell infiltration in upper dermis with focal basal layer vacuolar degeneration (**Figure 4**). These findings were consistent with the diagnosis of lichen planus.

Based on clinical findings and histopathology, patient was diagnosed as alopecia areata (ophiasis pattern) with segmental vitiligo and segmental lichen planus. Therapy was started with oral betamethasone in a dose of 0.1 mg/kg body weight twice a week. There was no significant improvement even after 4 months of therapy. Lichen planus lesions had somewhat flattened. However, vitiligo lesions and alopecia areata did not show any response

at all. We lost the patient to follow up thereafter.

Discussion

Alopecia areata (AA) is a chronic inflammatory condition that affects the hair follicles and sometimes the nails. Current evidence suggests that hair follicle inflammation in AA is caused by T cell-mediated autoimmune response in genetically predisposed persons.[1] There is an increased frequency of other autoimmune diseases, both cutaneous and extracutaneous conditions.[2] This discussion will focus on cutaneous associations only. Various dermatological conditions have been described in association of AA. Of them, vitiligo is most notable and is found in 1.8-3% cases of AA.[3] Vitiligo vulgaris was the most common type, followed by focal, acrofacial, segmental and universal types.[4] Localized vitiligo and segmental vitiligo have been rarely described in association with AA, in part due to low prevalence of these types.[5] Recently, vitiligo has been reported as a complication of diphencyprone therapy for AA.[6,7] Other associations include atopic dermatitis,[8] chronic mucocutaneous candidosis,[9] lichen planus,[10] scleroderma,[11,12] lichen sclerosus,[11] twenty nail dystrophy,[13] and morphea.[14] More recently, cutis verticis gyrata has been described in association with AA.[15]

As widely accepted, autoimmunity plays a key role in pathogenesis of AA, LP and vitiligo. Therefore, their co-occurrence may be explained by involvement CD8+ autoreactive T lymphocytes in pathogenesis of these conditions. The majority of the lymphocytes in the infiltrate of LP are CD8+ and CD45RO (memory)-positive cells and express the γ/δ T-cell-receptor.[16] The ensuing immune reaction by CD8+ T lymphocytes against activated keratinocytes results in epidermal cell damage and development of the lichenoid reaction that is the hallmark of lichen planus. Also, there is predominant accumulation of CD8+ T cells around hair follicles in alopecia areata with evidence of IFN-γ induced collapse of normal immune privilege of hair follicles.[17] Similarly, in vitiligo, an increased number of circulating CD8+ T lymphocytes reactive to Melan-A/Mart1, glycoprotein 100, and tyrosinase has been reported. Activated CD8+T cells have been demonstrated in perilesional vitiliginous skin.[18]

Our case was unique in having segmental vitiligo and segmental LP in association with AA. The segmental nature of vitiligo and LP may be explained by considering genetic predisposition for these conditions and mosaicism. However, this needs to be confirmed by further studies. The nail findings i.e. pterygium and longitudinal splitting cannot be explained by AA. These findings are suggestive of LP changes of nails, though histopathological confirmation was not done for lack of consent for nail biopsy. Both AA and LP changes of nails may coexist; though, it has not been reported frequently in literature. Moreover, poor treatment outcome in our patient with oral corticosteroid necessitates search for widely accepted and validated treatment for complicated "AA".

References

1. Messenger AG, Berker DAR, Sinclair RD. Disorders of hair. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rook's Textbook of Dermatology. 8th ed*. West Sussex: Wiley Blackwell; 2010. P. 66.31-7
2. Paus R, Olsen EA, Messenger AG. Hair growth disorders. In: Wolff K, Goldsmith LA, Katz SI *et al*., editors. *Fitzpatrick's Dermatology in General Medicine. 7th ed*. New York: McGraw-Hill; 2008. P. 762-5.
3. Bolduc C, Lui H, Shapiro J. Alopecia areata. [cited 2011July 15]. Available from: http://emedicine.medscape.com/article/1069931-overview

4. Akay BN, Bozkir M, Anadolu Y, Gullu S. Epidemiology of vitiligo, associated autoimmune diseases and audiological abnormalities: Ankara study of 80 patients in Turkey. *J Eur Acad Dermatol Venereol* 2010;**24**:1144-50.
5. Kuchabal SD, Kuchabal DS. Alopecia areata associated with localized vitiligo. *Case Rep Dermatol* 2010;**27**:27-31.
6. Henderson C A, Ilchyshyn A. Vitiligo complicating diphencyprone sensitization therapy for alopecia universalis. *Br J Dermatol* 1995;**133**:496-7.
7. Hatzis J, Gourgiotou K, Tosca A *et al.* Vitiligo as a reaction to topical treatment with diphencyprone. *Dermatologica* 1988; **177**:146-8.
8. Friedmann PS, Arden-Jones MR, Holden CA. Atopic dermatitis. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rook's Textbook of Dermatology*. *8th ed*. West Sussex: Wiley Blackwell Publishers; 2010. P. 24.23.1-23.24.
9. Hay RJ, Ashbee HR. Mycology. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rook's Textbook of Dermatology. 8th ed*. West Sussex: Wiley Blackwell Publishers; 2010. P. 36.1-36.94.
10. Breathnach SM. Lichen planus and lichenoid disorders. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rook's Textbook of Dermatology. 8th ed*. West Sussex: Wiley Blackwell Publishers; 2010. P. 41.1-41.28.
11. Goodfield MJD, Jones SK, Veale DJ. The 'Connective Tissue Diseases'. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rook's Textbook of Dermatology. 8th ed*. West Sussex: Wiley Blackwell Publishers; 2010. P. 51.1-51.138.
12. Brenner W, Diem E, Gschnait F. Coincidence of vitiligo, alopecia areata, onychodystrophy, localized scleroderma and lichen planus. *Dermatologica* 1979;**159**:356-60.
13. Berker DAR, Baran R. Disorders of Nail. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rook's Textbook of Dermatology*. *8th ed*. West Sussex: Wiley Blackwell Publishers; 2010. P. 65.1-65.57.
14. Poojary SA. Vitiligo and associated autoimmune disorders: A retrospective hospital-based study in Mumbai, India. *Allergol Immunopathol* (Madr) 2011. doi:10.1016/j.aller.2010.12.00 (In press)
15. Yoo KH, Lee JW, Jang WS *et al*. Cutis verticis gyrata and alopecia areata: a synchronous coincidence? *Yonsei Med J* 2010;**51**:612-4.
16. Gadenne AS, Strucke R, Dunn D *et al*. T-cell lines derived from lesional skin of lichen planus patients contain a distinctive population of T-cell receptor gamma delta-bearing cells. *J Invest Dermatol* 1994;**103**:347-51.
17. Gilhar A, Paus R, Kalish R. Lymphocytes, neuropeptides, and genes involved in alopecia areata. *J Clin Invest* 2007;**117**:2019-27.
18. Halder RM, Taliaferro SJ. Vitiligo. In: Wolff K, Goldsmith LA, Katz SI *et al.*, editors. *Fitzpatrick's Dermatology in General Medicine. 7th ed*. New York: McGraw-Hill; 2008. P. 616-22.