Poikiloderma Vasculare Atrophicans Showing Features of Ashy Dermatosis in the Beginning

Jiehyun Jeon, Joo Ha Kim, Jae Woo Ahn, Hae Jun Song

Department of Dermatology, Korea University Guro Hospital, Seoul, Korea

Poikiloderma vasculare atrophicans (PVA) is a rare poikilodermatous variant of early-stage mycosis fungoides characterized by generalized poikiloderma, atrophy, mottled dyspigmentation, and telangiectasia. In 2001, a 14-year-old male presented with asymptomatic brownish-gray polymorphic macules throughout the body with flexural accentuation. A skin biopsy showed increased melanophages with focal hydropic changes. Ashy dermatosis was considered a possible diagnosis. In 2005, the lesions began to show darkening and lichenification in the lower part of the trunk. In 2011, his skin showed definite poikilodermatosus changes, and a biopsy showed band-like inflammatory infiltrations of atypical lymphocytes, epidermal atrophy, and epidermotropism of predominantly CD4 − CD8 + atypical T cells. In addition, results of T-cell receptor gene rearrangement analysis were positive. Based on the aforementioned findings, he was diagnosed with PVA. If a patient shows long-standing and progressive hyperpigmentary skin changes, periodic follow-up and repeated skin biopsies are recommended to determine the underlying condition. (Ann Dermatol 27(2) 197 ~ 200, 2015)

Keywords - CD4, CD8, CD4-CD8 ratio, Mycosis fungoides, Poikiloderma, T-lymphocytes

INTRODUCTION

Ashy dermatosis is an idiopathic ash-colored macular hyperpigmentation disorder in young adults. It is usually slowly progressive and leaves permanent discoloration. Poikiloderma vasculare atrophicans (PVA) is poikilodermatosus mycosis fungoides (MF) characterized by poikilodermatosus hyper- and hypopigmentation, atrophy, and typical telangiectasia on the trunk. PVA with an epidermotropism of CD4 − CD8 + atypical T cells responds well to phototherapy and has a good overall prognosis. Herein, we present a case of PVA showing features of ash dermatosis in the beginning.

CASE REPORT

In 2001, a 14-year-old male patient presented with asymptomatic brownish black-pigmented reticulated patches over the whole body, particularly on the neck, axillary, and antecubital areas, for an unknown period (Fig. 1A). The results of laboratory studies were all within normal limits or were negative. The histologic examination showed increased number of melanophages in the papillary dermis with focal hydropic changes in the basement membrane zone (Fig. 2A). Ashy dermatosis was considered a possible diagnosis, but the patient was lost to follow-up. In 2005, the patient revisited with scaly increased darkening and lichenification on the previous lesions in the absence of any interim treatment (Fig. 1B). The skin biopsy revealed interface dermatitis with band-like infiltration in the upper dermis with focal hydropic changes in the basement membrane zone (Fig. 2A). Ashy dermatosis was considered a possible diagnosis, but the patient was lost to follow-up. In 2005, the patient revisited with scaly increased darkening and lichenification on the previous lesions in the absence of any interim treatment (Fig. 1B). The skin biopsy revealed interface dermatitis with band-like infiltration in the upper dermis with focal hydropic changes in the basement membrane zone (Fig. 2A). Ashy dermatosis was considered a possible diagnosis, but the patient was lost to follow-up.
Fig. 1. (A) Dusky, dark brownish pigmented reticulated patches over the whole body. (B) Scaly increased darkening and lichenification on the previous lesions. (C) Additional prominent poikilodermatous change in the previous skin lesions. (D) The poikilodermatous lesions have disappeared after narrow-band ultraviolet B phototherapy.

sions (Fig. 1C). The physical examination and laboratory test results were all within normal limits or were negative. A skin biopsy performed on the flank area showed epidermal atrophy and lymphocytic infiltration in the upper dermis with epidermotropism of atypical lymphocytes (Fig. 2C). A CD4~CD8 double stain revealed a 1:1 ratio of CD4 cells to CD8 cells, and the atypical T cells were mainly CD4−CD8+ (Fig. 2C'). A retrospective CD4-CD8 double staining on the previous biopsy specimen also revealed a 1:1 ratio of CD4 cells to CD8 cells and mild epidermotropism of CD4−CD8+ lymphocytes (Fig. 2A', B'). The results of the T-cell receptor (TCR) γ gene rearrangement analysis were positive for clonality in the affected skin lesions. Based on the aforementioned findings, PVA (stage IB) was diagnosed. We treated the generalized skin involvement with topical steroids and narrow-band ultraviolet B (NB-UVB) phototherapy. In March 2012 after 1 year of treatment, the poikilodermatous lesions had disappeared and atypical lymphocytes were not observed on a follow-up skin biopsy (Fig. 1D, 2D, 2D'). The results of another TCR γ gene rearrangement analysis were also negative. Currently, the patient undergoes regular observation after 6 months of biweekly NB-UVB phototherapy. No new suspicious skin lesions or palpable lymph nodes have been observed thus far.

DISCUSSION

PVA was first introduced by Jacobi in 1908 to describe a condition that was later regarded as a dermatomyositis. Recent review articles have reported PVA as poikilodermatous MF characterized by mottled pigmentation (hyper- and hypopigmentation), atrophy, and typical telangiectasia involving the major flexural areas and the trunk. Abbott et al. identified 49 patients who predominately had poikilodermatous MF. There was a slight predominance of
male patients 1.6:1 (30 of 49) with a median age of 44 years. Most patients (43 of 49) had an early disease stage (≤IIA) at diagnosis, and no patients had stage IV disease at presentation. The histopathologic features, in addition to those associated with classic MF, included atypical T lymphocyte infiltration and epidermal atrophy in the papillary dermis and epidermis, pigmented incontinence, and telangiectatic vessels. In PVA, a predominant CD4<sup>+</sup>CD8<sup>-</sup> phenotype was detected on immunohistochemistry (15/40; 38%) compared with classic MF, which had a CD8<sup>-</sup> phenotype in just 6.5% of cases according to previous studies. Our patient also revealed mild epidermotropism of the CD4<sup>-</sup>CD8<sup>-</sup> lymphocytes in retrospective CD4-CD8 double staining accompanied by similar clinical features of ashy dermatosis in the beginning (Fig. 2A', B'). Thus, careful pathologic evaluation can lead to early diagnosis of the disease or at least a differential diagnosis. Patients with PVA respond well to phototherapy, which is
the most commonly used first-line therapy, and they have a good overall prognosis. To the best of our knowledge, four cases involving a similar presentation were reported in the Korean literature as parapsoriasis variegata in 1979, as poikilodermatous MF in 1999, and as PVA in 1999 and 2011.

Ashy dermatosis or erythema dyschromicum perstans presents as asymptomatic confluent ashy colored macular hyperpigmentation that is idiopathic, slowly progressive, and permanently discoloring, and usually appears in young adults. The trunk and proximal extremities are more commonly involved, followed by the neck and face. The histopathologic features show dermal perivascular lymphocytic infiltration, vacuolization of the basal cell layer, occasional colloid bodies, and incontinence of pigment. Since there is no effective treatment, a number of different therapeutic approaches have been attempted. CD4−CD8+ MF is a rare form of MF (fewer than 5%), which is characterized by chronic epidermotropism of predominately CD8+ atypical T cells and often hyperpigmentation and poikiloderma. Nikolaou et al. presented a series of seven cases of CD8+ cytotoxic variant MF and reported that CD8+ MF follows an indolent course, responds well to phototherapy, and has a benign prognosis. Based on these findings, they suggested that the CD8+ immunophenotype may represent a marker of mild biological behavior. Our patient also showed a CD4−CD8+ immunophenotype, which may explain the benign clinical course over a 10-year period. Thus, if a patient shows long-standing and progressive hyperpigmentary skin changes, periodic follow-up and repeated skin biopsies are recommended to determine the underlying condition.

In conclusion, our case demonstrates four important points. First, the clinical and pathologic findings of early stage PVA can mimic ashy dermatosis. Therefore, if a patient shows long-standing and progressive hyperpigmentary skin changes, periodic follow-up and repeated skin biopsies are recommended to determine the underlying condition. Second, we confirmed that PVA or poikilodermatous MF is a rare variant of MF, which has a benign clinical course and presents with the CD8− phenotype more frequently. Third, we also confirmed that CD8+ MF presents with special clinical features such as hyperpigmentation and poikiloderma. Finally, this case had an indolent, benign course, which corroborates the finding that the CD8+ immunophenotype of MF may represent a marker of mild biologic behavior.

REFERENCES

1. Dowling GB, Freudenthal W. Dermatomyositis and poikiloderma atrophicans vascularis: a clinical and histological comparison. Br J Dermatol 1938;50:519-539.
2. Howard MS, Smoller BR. Mycosis fungoides: classic disease and variant presentations. Semin Cutan Med Surg 2000;19:91-99.
3. Kazakov DV, Burg G, Kempf W. Clinicopathological spectrum of mycosis fungoides. J Eur Acad Dermatol Venereol 2004;18:397-415.
4. Abbott RA, Sahni D, Robson A, Agar N, Whittaker S, Scarsbrick JJ. Poikilodermatous mycosis fungoides: a study of its clinicopathological, immunophenotypic, and prognostic features. J Am Acad Dermatol 2011;65:313-319.
5. Willemze R, Jaffe ES, Burg G, Cerroni L, Berti E, Swerdlow SH, et al. WHO-EORTC classification for cutaneous lymphomas. Blood 2005;105:3768-3785.
6. McKee PH, Calonje E, Granter SR. Pathology of the skin with clinical correlations. Philadelphia: Elsevier, 2005.
7. Nikolaou VA, Papadavid E, Katsambas A, Stragitis AJ, Marinou I, Anagnostou D, et al. Clinical characteristics and course of CD8+ cytotoxic variant of mycosis fungoides: a case series of seven patients. Br J Dermatol 2009;161:826-830.
8. Lee JB, Myung KB, Kim JH. Parapsoriasis variegata: report of a case. Korean J Dermatol 1979;17:367-371.
9. Jang HC, Ahn BJ, Kim SW, Kim DS. Case of poikilodermatous mycosis fungoides. Korean J Dermatol 1999;37:1515-1517.
10. Cho KH, Lee HS. A clinicopathological study of early mycosis fungoides. Korean J Dermatol 1999;37:838-845.
11. Choi MS, Lee JB, Kim SJ, Lee SC, Won YH, Yun SJ. A case of poikilodermatous mycosis fungoides. Ann Dermatol 2011;23(Suppl 1):S48-S52.
12. Torrelo A, Zaballos P, Colmenero I, Mediero IG, de Prada I, Zambrano A. Erythema dyschromicum perstans in children: a report of 14 cases. J Eur Acad Dermatol Venereol 2005;19:422-426.