J Korean Med Sci 2000; 15: 724-6
ISSN 1011-8934

Macular Amyloidosis Presented as Poikiloderma : A Case Report

We report a 51-year-old Vietam War veteran with an unusual variant of macular amyloidosis presenting as poikilodermatous skin lesions. The extensive mottled brown pigmentation was checkered with small hypopigmented or normal skin-colored spots and intermingled with telangiectasia. Skin biopsy revealed subepidermal amyloid deposits. There was no evidence of extracutaneous involvements. This case could be easily confused with other true poikiloderma lesions.

Key Words: *Macular Amyloidosis; Poikiloderma*

Hee Young Kang, Won Hyoung Kang

Department of Dermatology, Ajou University School of Medicine, Suwon, Korea

Received: 8 November 1999
Accepted: 13 December 1999

Address for correspondence
Won Hyoung Kang, M.D.
Department of Dermatology, Ajou University School of Medicine, 5 Wonchon-dong, Paldal-gu, Suwon 442-721, Korea
Tel: +82.31-219-5191, Fax: +82.31-216-9189
E-mail: wwww@madang.ajou.ac.kr

INTRODUCTION

Primary cutaneous amyloidosis is characterized by the deposition of amyloid in normal skin with no evidence of systemic involvement. There are three well-known clinical variants: macular, lichenoid and nodular amyloidosis (1). They are easily distinguished by their clinical features, i.e. macular amyloidosis is characterized by rippled or reticulated pigmented macules on the upper back or limbs and lichenoid amyloidosis manifests as pruritic, pigmented, lichenoid papules on the extensor aspects of extremities. We report a patient with an unusual variant of macular amyloidosis who had the clinical appearance of a poikiloderma and showed subepidermal amyloid deposits in histopathologic examination.

CASE REPORT

A 51-year-old Korean man presented with asymptomatic generalized hyperpigmentation for more than 20 years. He had participated in the Vietnam War during 1971-1972. One year after his return from the war, he noticed hyperpigmented skin lesions. He insisted that his skin lesions has been developed after exposure to Agent Orange (a defoliant used in the Vietnam War). He was otherwise in good health. Physical examination revealed an extensive, mottled, brown pigmented macules or patches distributed over the back, abdomen and limbs. The pigmentation was checkered with small hypopigmented or normal skin-colored spots and intermingled with telangiectasia (Fig. 1). The skin was dry with an atrophic appearance. There was no discrete papular component. Our first impression was poikiloderma atrophica vasculare and a 2 mm punch biopsy was done from two different sites, sun-exposed and -covered areas.

Skin samples from the hyperpigmented macules of two

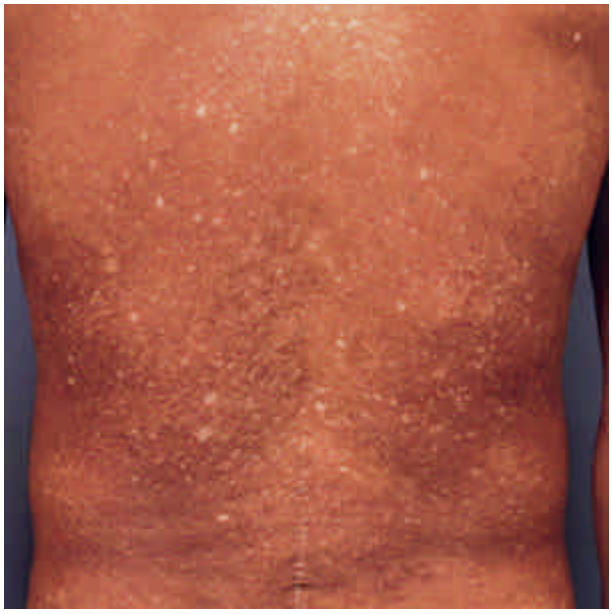

Fig. 1. Mottled hyperpigmentation checkered with hypopigmented or normal skin-colored spots and intermingled with telangiectasia.

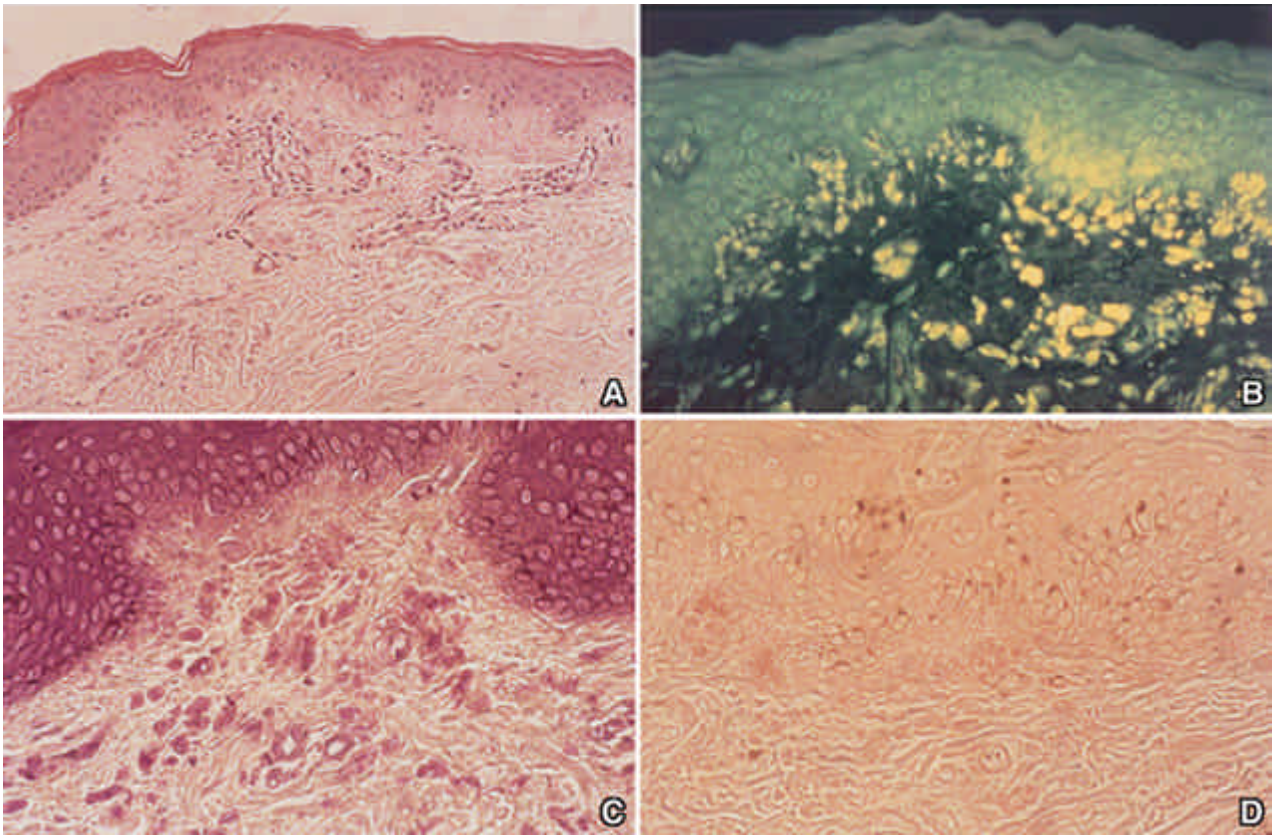

Fig. 2. Histopathologic findings of skin lesion from the back. There were telangiectasia and globular deposits of pale pink amorphous substances in the upper dermis (**A**: H&E, ×100). These deposits showed up as bright yellow fluorescence under polarizing microscopy when stained with thioflavin T (**B**: thioflavin T, ×400) and metachromasia when stained with crystal violet (**C**: crystal violet, ×400). There was weak positive staining with Congo red (**D**: Congo red, ×400).

different lesions, the back and the left forearm, demonstrated the same histopathologic features; telangiectasia and globular deposits of pale pink amorphous substances in the upper dermis (Fig. 2A). These deposits showed up as bright yellow fluorescence under polarizing microscopy with thioflavin T (Fig. 2B) and stained metachromatically purple with crystal violet (Fig. 2C). There was weak positive staining with Congo red (Fig. 2D) and negative apple-green birefringence on polarized light examination.

Based on the clinical picture of poikiloderma and the histology of cutaneous amyloidosis, we made the diagnosis of primary cutaneous amyloidosis (macular type) presented as poikiloderma. No treatment was given.

DISCUSSION

Lichen amyloidosis presenting as poikilodermatous skin lesions has been rarely reported (2-4). Marchionini et al. (2) reported an elderly patient who had lichenoid amyloidosis associated with poikilodermatous skin lesions and entitled it as 'lichenoid and poikiloderma-like amyloidosis'. Later, poikiloderma-like cutaneous amyloidosis (PCA) syndrome was added to the classification as a rare variant of primary cutaneous amyloidosis (3, 4). The PCA syndrome includes lichenoid papules, histological evidence of amyloid deposits and poikilodermatous skin manifestations which may appear early in life. Light sensitivity, short stature, palmoplantar hyperkeratosis and blister formations are frequently associated. In our patient, the macular pattern of the lesions did not progress to lichenoid papules over a period of 20 years. So we consider our patient to have a pure macular amyloidosis presented as poikiloderma, which was first reported by Serna-Perez et al. (5). The poikilodermatous pattern shown by our patient can be easily misdiagnosed as other true poikiloderma which shows no amyloid deposits on histological examinations.

The herbicide Agent Orange [a 1:1 mixture of 2,4-dichlorophenoxyacetic acid and 2,4,5-trichlorophenoxyacetic acid in diesel oil] was sprayed during the Vietnam War. The herbicide was contaminated with dioxins. Since the late 1970s, several epidemiologic studies have appeared linking exposure to dioxin to some skin lesions

(6). Chloracne is the most sensitive indicator of significant dioxin exposure. Porphyria cutanea tarda and hyperpigmentation are other known cutaneous effects, and malignant fibrous histiocytomas of the skin may possibly be associated. The cause of macular amyloidosis is not clear. Prolonged friction from a rough nylon towel or a back scratcher is thought to be cause of macular amyloidosis. To date, there has been no report that exposure to dioxins was related to either poikiloderma or amyloidosis. At present we can not conclude the association between dioxins and the skin lesions in our patient. It is unclear whether it is a coincidence or a causal relationship.

Histologically, amyloid deposits are recognized as eosinophilic globules in the upper dermis on H&E staining. They can be identified by Congo red, methyl or crystal violet, triphenyl-methane dyes and thioflavin T staining methods (7). We performed crystal violet, thioflavin T and Congo red stains on our specimen. In our study, the purple staining of amyloid by crystal violet or fluorescence by thioflavin T was more definite and informative than with Congo red stains. With Congo red, the amyloid deposits stained weakly and showed no birefringence (apple-green) in polarizing microscopy. These results are consistent with other reports; Hashimoto et al. (8) showed variable staining properties of Congo red and Ho et al. (4) reported weak or negative stainings with Congo red and positive stainings with crystal violet.

In conclusion, we present this unusual case to alert dermatologists that macular amyloidosis can manifest itself as poikilodermatous skin lesion. We recommend that skin biopsies should be conducted in order to confirm the diagnosis.

REFERENCES

1. Wang WJ. *Clinical features of cutaneous amyloidoses. Clin Dermatol 1990; 8: 13-9.*
2. Marchionini A, John F. *Ueber lichenoide und poikilodermieartige Haut-amyloidose. Arch Dermatol Syphilol 1936; 173: 545-61.*
3. Ogino A, Tanaka S. *Poikiloderma-like cutaneous amyloidosis. Report of a case and review of literature. Dermatologica 1977; 155: 301-9.*
4. Ho MH, Chong LY. *Poikiloderma-like cutaneous amyloidosis in an ethnic Chinese girl. J Dermatol 1998; 25: 730-4.*
5. Serna-Perez MJ, Vazquez-Doval FJ, Idoate M, Sola Casas MA, Quintanilla E. *Extensive macular amyloidosis associated with poikiloderma. Int J Dermatol 1992; 31: 277-8.*
6. Dunagin WG. *Cutaneous signs of systemic toxicity due to dioxins and related chemicals. J Am Acad Dermatol 1984; 10; 688-700.*
7. Thewes M, Hein R, Ring D. *Lichen amyloidosus. Eur J Dermatol 1999; 9: 63-4.*
8. Hashimoto K, Gross BG, Lever WF. *Lichen amyloidosis. Histochemical and electron microscopic studies. J Invest Dermatol 1965; 65; 204-19.*