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A Case of Benign Atrophic Papulosis in a Young Male

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Dear Editor:

Atrophic papulosis (Degos disease) is a rare, thrombo-occlusive, multisystem disease. However, some patients have been described with a benign clinical course of sole skin involvement[1]. Herein, we present a 32-year-old male patient with classic skin lesions without systemic involvement during 7 years' follow-up.

A 32-year-old male was admitted to our department for a one-year history of multiple erythematous papules with atrophic porcelain-white center on the trunk and extremities. He denied a history of headaches, arthritis, visual symptoms, gastrointestinal discomfort or any constitutional symptoms. Physical examination revealed numerous 3~10 mm papules with a porcelain-white atrophic center and an erythematous halo, alongside scattered atrophic white scars (Fig. 1A, B). The lesions were distributed across his trunk and limbs.

Results of routine laboratory tests were all normal. His neu-

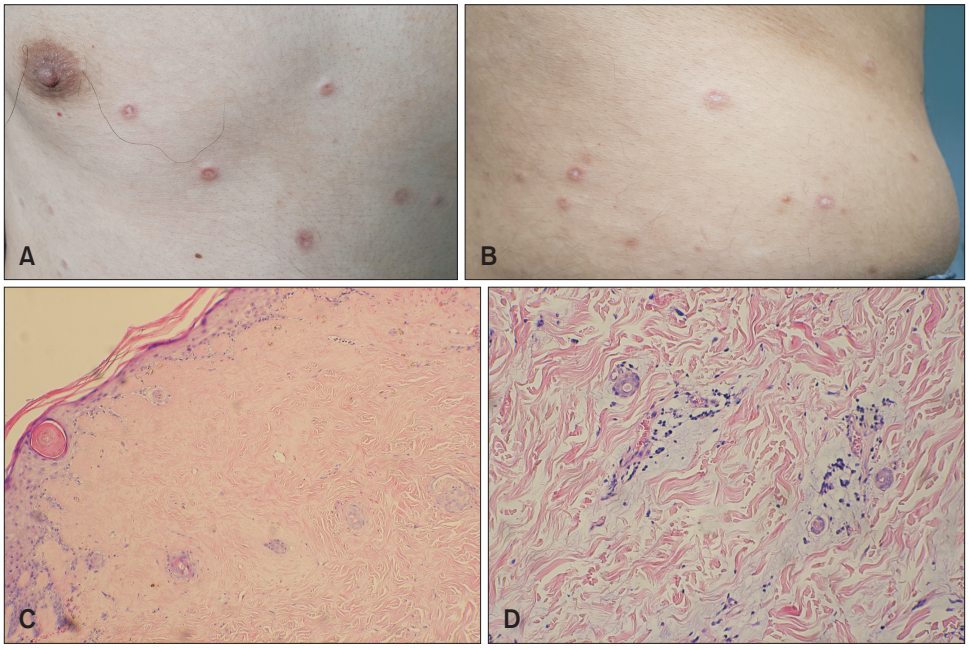

Fig. 1. (A, B) Clinical photographs before treatment. (A) Papules on the chest with white atrophic center and an erythematous rim. (B) Lesions on the back similar to those of the chest, alongside atrophic white scars. (C, D) Histopathological examination (C: H&E, original magnification ×40; D: H&E, original magnification ×200): back biopsy shows hyperkeratosis and epidermis atrophy, perivascular lymphocytic infiltration, melanin incontinence, homogeneous degeneration of collagen and wedge-shaped necrosis in dermis. There is also striking mucin deposition within the dermis.

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rological and mental state examination had unremarkable results. Electrocardiography, echocardiography, and abdominal ultrasonography showed no abnormalities. Histopathological examination showed hyperkeratosis and epidermis atrophy, perivascular lymphocytic infiltration, melanin incontinence, homogeneous degeneration of collagen and wedge-shaped necrosis in dermis. There was also striking mucin deposition within the dermis (Fig. 1C, D). The diagnosis of Degos disease (benign atrophic papulosis [BAP]) was made.

Treatment was started with dipyridamole 25 mg twice a day. The color of rash became lighter after 5 months. Treatment discontinued after two years. After five years, the lesions continued to spread on his trunk and extremities. He reported no systemic symptoms to date. A Longer follow-up is still needed.

In 2014, Zouboulis et al. proposed that the previously so-called malignant atrophic papulosis (MAP) should be renamed as atrophic papulosis which could be classified into two variants: MAP and BAP[1,2]. It usually occurs between the 20th~50th year of life[1]. Pediatric cases have also been reported[3]. The differences between MAP and BAP is shown in the Table 1[1-4]. MAP and BAP can be distinguished only by the presence of systemic disease in MAP, which has a median onset of 1 year (0.03~0.97 quantiles: 0~7 years) after cutaneous presentation and portends a poor prognosis[1]. The probability of a benign course increased with time, from 71% at onset to 97% after 7 years of monosymptomatic cutaneous disease[1]. The etiology of atrophic papulosis remains unknown but coagulopathy, vasculitis and primary endothelial dysfunction are the most commonly suggested hypotheses[1]. Recently, anomalies in complement activation C5b-9 and dysregulation of interferon-α have been implicated[5]. Histopathologic findings include epidermal atrophy, dermal perivascular lymphocytic infiltrate, interstitial mucin deposition, and wedge-shaped dermal necrosis[1], which are coincident with ours. Treatment options for atrophic papulosis are limited. First line treatments include aspirin, pentoxifylline, dipyridamole, ticlopidine. Recently, treatment with eculizumab and treprostinil have demonstrated partial improvement[4]. When systemic involvements such as acute abdomen or cerebral artery thrombosis occurs, surgical operation and thrombolytic therapy are feasible.

In summary, we present a benign variant of atrophic papulosis without systemic involvement during 7-years' follow-up, emphasizing the different course between clinical spectrum of malignant variant and benign form. Because systemic involvement may develop years after the onset of cutaneous lesions, it is crucial to perform regular follow-up of the patients.

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We received the patient's consent form about publishing all photographic materials.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

Table 1. Different characteristics between malignant atrophic papulosis (MAP) and benign atrophic papulosis (BAP)

Characteristic	MAP	BAP
Sites involved	Gastrointestinal tract, pericardial cavity, pleural cavity, muscles, diaphragm, central nervous system, myocardium, lungs, eyes, skin	Only skin
Cutaneous lesions	A depressed atrophic plaque with a white hue and a peripheral violaceous rim	
Vulnerable population	In the third to fifth decade of life	
	Male	Female
Histopathologic features	A superficial and deep perivascular and periadnexal lymphocytic infiltrate that is accompanied by interstitial mucin deposition. Inverted, wedge-shaped dermal necrosis	
Treatment	Heparin and antiaggregant (or platelet aggregation-inhibiting) drugs, eculizumab and treprostinil	
	When bowel perforation or cerebral artery thrombosis occurs, surgical operation and thrombolytic therapy are feasible	-
Prognosis	Cumulative 5-year survival rate of MAP is approximately 55.4%	No patient had a lethal outcome

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