

Anosacral amyloidosis in a Chinese-Caribbean male



Caroline A. Gerhardt, BS,^a Brandon Cardon, MD,^b Paul Rodriguez-Waitkus, MD, PhD,^b
Lucia Seminario-Vidal, MD, PhD,^b and Wei-Shen Chen, MD, PhD^b
Tampa, Florida

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INTRODUCTION

Anosacral amyloidosis (ASA) is a rare type of primary cutaneous amyloidosis that has only been described in Asian populations and countries.¹ The lesions present as well-demarcated, brown-colored patches or plaques, fanning out in lines from the anus to the sacral region. This presentation was first reported under the term “hyperkeratotic lichenified skin of the gluteal region” in 1979 in Japan.² In 1981, a biopsy of the gluteal skin demonstrated amyloid deposition, and the diagnosis term of ASA was coined.³ Since then, additional cases of similar gluteal lesions have been reported in Japan, China, and Taiwan.⁴⁻⁶ Here, we report a case of ASA in a Chinese-Caribbean man. To our knowledge, this is the first reported case in the United States.

CASE REPORT

A 75-year-old man of Chinese-Caribbean descent presented for a routine total-body skin examination. On examination, brown, erythematous, scaly plaques were noted in a fan-like distribution on the gluteal cleft region (Fig 1, A and B). The patient reported that the skin changes had been present for several years and denied any symptoms such as pruritus or pain in the region.

The clinical differential diagnosis at presentation included psoriasis, lichen simplex chronicus, and mycosis fungoides. A 4-mm punch biopsy was taken from the left gluteal cleft and demonstrated basal keratinocyte hyperpigmentation, increased melanophages in the superficial perivascular distribution, and a collection of eosinophilic homogenous material in the papillary dermis (Fig 2, A and B). These

Abbreviations used:

ASA: anosacral amyloidosis
SGD: senile gluteal dermatoses

amorphous aggregates were highlighted on pan-cytokeratin immunostain and Congo red special stain and demonstrated apple-green birefringence under a polarizing microscope (Fig 2, C-E). No fungal infection was identified on hematoxylin-eosin stain or by periodic acid–Schiff special stain (not shown). These histologic findings were compatible with lichen amyloidosis. At this anatomic location, the diagnosis of ASA was rendered.

Treatment with 0.1% triamcinolone cream was recommended. However, as the condition is benign and was asymptomatic, the patient deferred treatment.

DISCUSSION

ASA typically presents as well-demarcated, brown-colored patches and plaques, fanning out in lines from the anus to the sacral region. The presentation of ASA can vary based on severity. Mild cases of ASA show only slight lichenification with light brown color, while severe cases show dark brown color and obvious hyperkeratosis.⁴ Many patients with ASA also have concurrent lesions of either macular or lichenoid amyloidosis on other parts of their bodies.^{4,5} However, our patient did not appear to have any other involved body sites. While the etiology of ASA has not been elucidated, the likely cause of the common types of primary cutaneous

From the University of South Florida Morsani College of Medicine^a; and the Department of Dermatology & Cutaneous Surgery, University of South Florida.^b

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Correspondence to: Wei-Shen Chen, MD, PhD, 12901 Bruce B. Downs Boulevard, 4th Floor, Tampa, FL 33612. E-mail: weishenchen@usf.edu.

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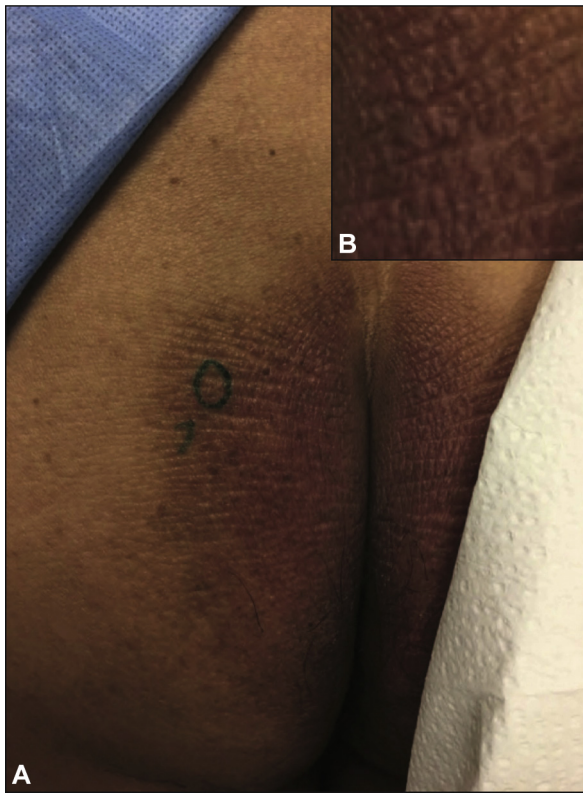


Fig 1. Well-circumscribed, brown, erythematous, scaly plaque in the gluteal cleft region (**A**). High-power view emphasizing the horizontally oriented, lichenified ridges (inset, **B**).

amyloidosis is friction from scratching.⁵ In the reported cases of ASA, many of the patients experienced pruritus of the involved area; however, it is not clear if the pruritus preceded the cutaneous changes.⁴ Interestingly, our patient denied pruritus, history of scratching, and friction to the affected area.

Histologically, ASA is characterized by hyperkeratosis, an increase in melanin in the basal layer, pigment incontinence, and amyloid deposition within the papillary dermis. The proposed mechanism causing primary cutaneous amyloidosis is the filamentous degeneration theory. This theory hypothesizes that degenerating epidermal cells, triggered by apoptosis, are discharged into the dermis and later converted into amyloid.^{7,8} Wang et al's⁴ case series supported this mechanism, as many of their cases demonstrated dyskeratosis and vacuolar alteration of the basal cell, which may indicate the occurrence of apoptosis.

The treatment of ASA can be challenging. There has been reported beneficial use of topical corticosteroids, oral retinoids, and cyclophosphamide.¹ Topical corticosteroids may help stabilize the disease

process; however, the resolution of ASA has not been reported.⁴ Topical corticosteroids may also be helpful for patients experiencing pruritus.

ASA should be distinguished from senile gluteal dermatoses (SGD). Originally, these 2 disease processes were considered to be of the same entity due to their similar clinical presentation, in primarily Asian patients. However, more recently, after much controversy, the 2 have been distinguished from one another based on histologic findings. Characteristic clinical features of SGD consist of hyperkeratotic, lichenified, brownish patches or plaques exhibiting fine hyperkeratotic ridges.^{5,6,9} These lesions have been reported both at the anal cleft and at the skin overlying the coccygeal apex as well as the ischial tuberosities—a distribution known as “3 corners of a triangle.”⁵ Histopathology typically demonstrates hyperkeratosis, acanthosis, follicular plugging, and vascular dilation in the papillary dermis, with minimal lymphohistiocytic infiltrate.^{5,9,10} Notably, amyloid is absent in SGD.

The proposed etiology of SGD is related to mechanical irritation due to prolonged sitting on hard surfaces. Moon et al⁹ found that SGD is significantly associated with prolonged daytime sitting (>8 hours) and the use of a Korean-style mattress (thin cotton pad placed on a traditional hard flooring with a heating system). In Japan, a country with some of the most frequently reported cases, there is a custom of sitting on “tatami,” straw mats used as flooring.⁶ Cultural practices appear as a possible explanation for the prevalence of SGD in Asia.^{6,9}

The treatment of SGD with topical steroids and keratolytics have yielded unsatisfactory results.⁹ However, topical corticosteroids may help relieve the associated pruritus. Topical retinoids may help normalize epithelial differentiation and desquamation and have been beneficial for some patients.⁹ Most importantly, the prevention of SGD through pressure-relieving devices and avoidance of long periods of sitting has been helpful.^{9,10}

It is essential to acknowledge the differences between ASA and SGD due to the initial conflation. Other clinical differential diagnoses in this anatomic location may include inverse psoriasis and porokeratosis *ptychotropica*. However, the histologic findings would be distinct enough to not to be confused with ASA.

Given that ASA predominantly affects patients of Asian descent, this diagnosis has been mostly reported in Asian literature. Our patient is of Chinese descent; however, he grew up in the Caribbean and later moved to the United States. In the era of globalization and increased international migration, we are more

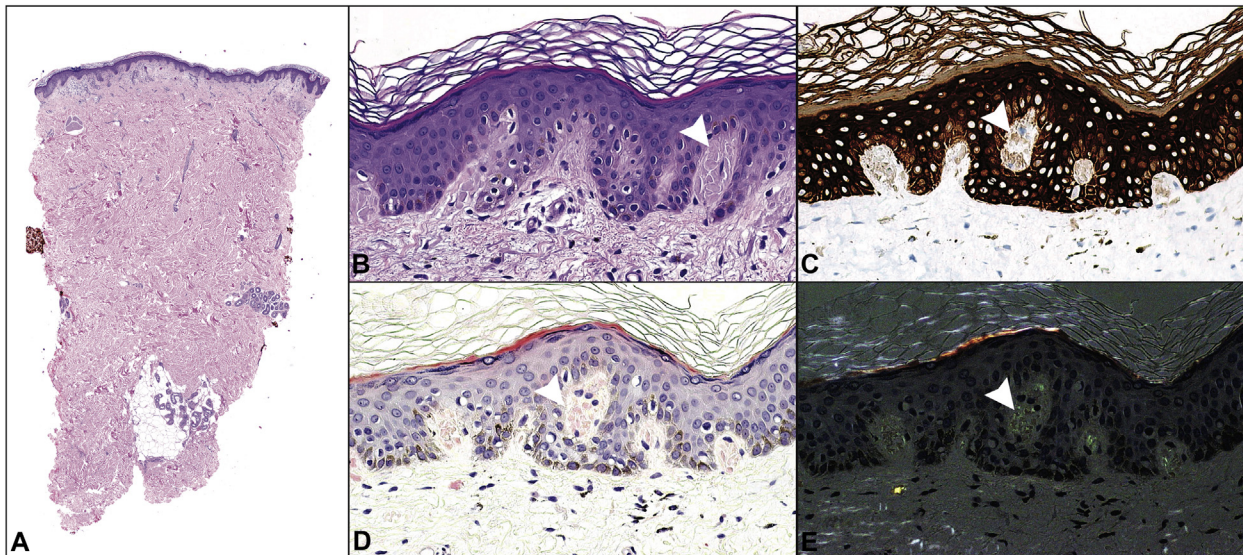


Fig 2. Overview of the biopsy from the gluteal cleft lesion, diagnosed as anosacral amyloidosis (A, Hematoxylin-eosin stain; original magnifications: $\times 20$). Basal keratinocyte hyperpigmentation, scattered melanophages in the superficial perivascular distribution, and a collection of eosinophilic homogenous amyloid granules within the papillary dermis (B, Hematoxylin-eosin stain; original magnifications: $\times 200$, *arrowhead*), which are highlighted by pan-cytokeratin staining (C, Hematoxylin-eosin stain; original magnifications: $\times 200$, *arrowhead*). These amyloid granules in the papillary dermis are highlighted by Congo red staining (D, Hematoxylin-eosin stain; original magnifications: $\times 200$, *arrowhead*) and demonstrate apple-green birefringence under a polarizing microscope (E, Hematoxylin-eosin stain; original magnifications: $\times 200$, *arrowhead*).

and more likely to encounter patients of international origin or with complex ethnic backgrounds. Herein, we describe to our knowledge the first reported case of ASA in the United States to acquaint our fellow colleagues with this unique disease for accurate diagnosis and proper management.

Conflicts of interest

None disclosed.

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