Mucinous Nevus

Min Young Lee, Ji Yeon Byun, Hae Young Choi, You Won Choi

Department of Dermatology, Ewha Womans University College of Medicine, Seoul, Korea

Mucinous nevus is an uncommon entity classified as either a cutaneous mucinosis or a connective tissue nevus. The condition presents as grouped papules and coalescent plaques growing in a unilateral or zosteriform manner. The key histopathological feature is a band-like deposition of mucin in the superficial dermis. A 34-year-old male presented with grouped gray-brown papules and confluent plaques exhibiting a zosteriform distribution on the right side of the lower back. The lesions had commenced in childhood. Histological examination revealed mucin deposition in the papillary dermis. Thus, we diagnosed a mucinous nevus. To date, only a few reports of such nevi have been reported in the literature. Therefore we report a rare case of mucinous nevus. (Ann Dermatol 30(4) 465 ~ 467, 2018)

-Keywords-Cutaneous, Mucin, Nevus

INTRODUCTION

Mucinous nevus is a rare disorder classified as either a cutaneous mucinosis or a connective tissue nevus^{1,2}. The condition was first described by Redondo Bellón et al.¹ in 1993. Clinically, asymptomatic grouped papules or pla-



ques grow to form a verrucous or nevoid feature exhibiting a unilateral or zosteriform distribution^{3,4}. The nevus usually develops on the trunk at birth or in early adulthood3,4. Histologically, the nevus is characterized by mucin deposits localized to the superficial dermis^{3,4}.

To the best of our knowledge, only 25 cases have been reported in the English-language literature^{3,5-11} and only one case in the Korean literature¹². Herein, we report an unusual case of mucinous nevus.

CASE REPORT

A previously healthy 34-year-old Korean male presented with asymptomatic grouped gray-brown papules and confluent plaques exhibiting a zosteriform distribution on his right lower back (Fig. 1). The skin lesions had commenced in childhood, gradually coalesced, and grew slowly. In our patient, pigmentary abnormalities as like freckles except for skin lesions of the right lower back were not observed. He had neither any past medical problem nor a family history of similar lesions and pigmentary abnormalities. He denied any trauma.

Histological examination revealed an acanthosis with elongated rete ridges and amorphous materials associated with loosely separated collagen fibers in the papillary dermis (Fig. 2A). The amorphous materials stained with alcian blue at pH 2.5 (Fig. 2B); this confirmed a mucin deposit limited to the papillary dermis. Verhoeff-van Gieson staining revealed that the numbers of elastic fibers in the papillary dermis were reduced in the regions of mucin deposition (Fig. 2C).

This clinicopathological analysis enabled us to diagnose a mucinous nevus. Our patient decided to allow us to observe the lesion; no treatment was performed.

DISCUSSION

Mucinous nevus is an uncommon entity initially described

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Corresponding author: You Won Choi, Department of Dermatology, Ewha Womans University School of Medicine, 1071 Anyangcheon-ro, Yangcheon-gu, Seoul 07985, Korea. Tel: 82-2-2650-2665, Fax: 82-2-2652-6925, E-mail: uwon313@ewha.ac.kr ORCID: https://orcid.org/0000-0001-6315-3889

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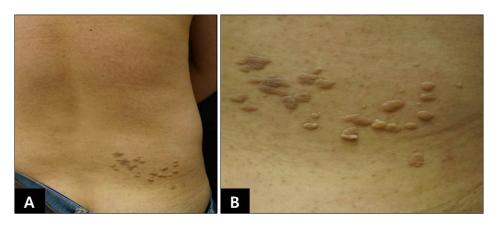


Fig. 1. (A) Multiple gray-brownish papules and confluent plaques with a zosteriform distribution on the right lower back. (B) Close-up view.

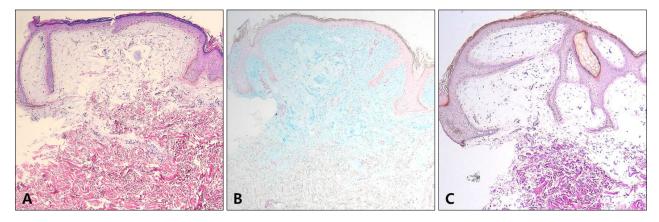


Fig. 2. (A) Amorphous materials and loosely separated collagen fibers in the papillary dermis and acanthosis with elongated rete ridges (H&E, \times 40). (B) Positively stained bluish amorphous materials in the papillary dermis (Alcian blue at pH 2.5, \times 40). (C) Reduced numbers of elastic fibers in the papillary dermis bearing mucin deposits (Verhoeff-van Gieson, \times 40).

by Redondo Bellón et al.¹ in 1993 and classified as either a cutaneous mucinosis or a connective tissue nevus^{1,2}. The cutaneous mucinoses are a heterogeneous group of diseases in which abnormal amounts of mucin are deposited in the skin⁶. Connective tissue nevi are hamartomas with unusual levels (excesses or deficiencies) of collagen, elastin, and/or proteoglycans².

The term "mucinous nevus" refers to the its nevoid appearance and the characteristic pattern of mucin deposits in the papillary dermis^{2,6,13}. Clinically, mucinous nevi present as asymptomatic, multiple skin-colored to brownish papules or plaques; separate lesions coalesce and then grow to form a verrucous or nevoid feature with a unilateral or zosteriform pattern^{3,4,6}. It usually develops at birth or in early adulthood^{6,13}. The principal site is the trunk, including the back^{3,6}. The male:female ratio is 5:1; the reason is not clear⁵. To date, there are two reports of familial mucinous nevus^{6,13}. However, there was no report about the genetic abnormality as like mosaicism. Histologically, mucinous nevus is characterized by diffuse band-like mucin deposits in the uppermost portion of the dermis¹⁻⁴. The mucin is thought to be composed of hyaluronic acid staining positively with alcian blue at pH 2.5 but not staining at pH 0.5^{14,15}. The origin of the mucin remains unknown¹⁴, but is presumed to be attributable to a primary metabolic process (such as overproduction) rather than a secondary catabolic process². Mucinous nevi are divided into two histopathological types depending on whether epidermal changes are present; these are connective tissue nevi of the proteoglycan (CTNP) type and the combined epidermal-CTNP type⁵. The epidermis is normal, in the CTNP type but, in the combined epidermal-CTNP type, exhibits hyperkeratosis and acanthosis with elongation of the rete ridges². Our case featured an epidermal change; thus, we diagnosed the combined epidermal-CTNP type of mucinous nevus.

Both an epidermal nevus and nevus lipomatosus superficialis exhibit nevoid features similar to those of a mucinous nevus. It is difficult, therefore, to clinically distinguish among the conditions. Histological data are necessary. Histologically, an epidermal nevus and nevus lipomatosus superficialis can be distinguished from a mucinous nevus; only the latter exhibits mucin deposits in the papillary dermis.

However, such mucin deposition is also observed in cutaneous mucinosis of infancy, but is very superficial and appears to be hugged by the epidermis⁵. Clinically, cutaneous mucinosis of infancy presents as scattered small papules unlike mucinous nevus.

Mucinous nevi do not require treatment (except for cosmetic purposes); the nevi are benign³. Surgical excision, scalpel dermabrasion, and carbon dioxide laser treatment are possible⁵. Surgical excision is not indicated if several discrete lesions are evident⁵. Mucinous nevus of the CTNP type was treated via scalpel dermabrasion, but scarring developed 1 year later¹⁶. There is one report of mucinous nevus of the combined epidermal-CTNP type which did not recur after carbon dioxide laser vaporization⁵. Our patient did not voice any cosmetic concern; thus, we decided to simply observe the lesions.

To the best of our knowledge, only 25 cases of mucinous nevi have been reported in the English-language literature^{3,5-11} and only one in the Korean literature¹². The principal location is the trunk including the back. Approximately 80% of all cases were reported in males and about 30% of all cases present at birth.

Herein, we report a rare case of mucinous nevus of the combined epidermal-CTNP type.

CONFLICT OF INTEREST

The authors have nothing to disclose.

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