Mucinous Nevus
-A Case Report and Review of Literature

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Mucinous nevus is a very rare entity and can be classified as both a cutaneous mucinosis and a connective tissue nevus. We describe the clinicopathologic features of an unusual case of mucinous nevus in a 9-year-old Taiwanese boy who presented with multiple grouped flesh-colored nodules with band-like distribution on his right thigh since birth. Histopathological examination of the skin lesion showed alcian blue (pH 2.5) and toluidine blue (pH 2.5) positive mucin deposited between the collagen bundles of the reticular dermis. (Dermatol Sinica 25: 147-152, 2007)

Key words : Mucinous nevus, Cutaneous mucinosis, Connective tissue nevus

黏液素母斑是一種非常罕見的疾病，可被歸類於皮膚黏液素病或結締組織母斑。在本文中，我們報告一個9歲台籍男孩，自出生即被發現多發性群聚性膚色結節成帶狀分佈於右大腿，組織病理學下可見有大量alcian blue(pH2.5) 及toluidine blue染色陽性的黏液沉積於網狀真皮層的膠原蛋白之間。(中華皮誌 25: 147-152, 2007)
INTRODUCTION

In 1993 Redondo Bellon et al. first described mucinous nevus (MN) in a 16-year-old female who presented with congenital, moderately pigmented plaque in the interscapular region. The biopsy specimen showed superficial dermal mucin deposition. So far 10 cases have been identified as MN in English literature. Some of them are congenital while others are acquired. We described a 9-year-old Taiwanese boy who presented with multiple, grouped flesh-colored nodules with band-like distribution on his right thigh since birth. The most important histopathologic feature was the presence of dermal mucin. We believe that our patient fall into the diagnosis of MN. To our knowledge, this is the first case identified in Taiwan.

CASE REPORT

A 9-year-old boy had multiple, asymptomatic, grouped flesh-colored nodules on his right thigh since birth. These nodules progressively increased in size in proportion with his growth. The patient is otherwise healthy. Family history is unremarkable. He did not receive any specific treatment before coming to our clinic. Physical examination revealed multiple, grouped flesh-colored nodules with band-like distribution on his right thigh. The surfaces of these lesions are smooth and the size of each nodule is 6-7 mm. The total distribution area is about 5×10 cm². (Fig. 1) A biopsy specimen showed basket-weave hyperkeratosis in the epidermis. In the reticular dermis, the collagen bundles were widely spaced. Alcian blue stain at pH 2.5 and toluidine blue stain both showed focally increased amounts of mucin in the reticular dermis. (Fig. 2, 3) Mucicarmine stain and periodic-acid Schiff stain revealed negative

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Fig. 1
Multiple flesh-colored nodules with band-like distribution on his right thigh.

Fig. 2
In this picture, right side is lesion part while left side is normal skin. Mucin deposit in reticular dermis stained positively with alcian blue (pH 2.5) (magnification x1); The amount of mucin is significantly increased in lesion part.

Fig. 3
Dermal mucin stains metachromatically purple with toluidine blue stain in the upper reticular dermis. (magnification x10).
finding. The number of fibroblasts in the lesion was increased (9 per high power field), comparing with normal skin (4 per high power field).

**DISCUSSION**

According to the clinical presentation and the pathologic findings of this case, all kinds of primary focal cutaneous mucinosis should be included in the differential diagnosis. Since mucicarmine stain and periodic-acid Schiff stain revealed negative finding, epithelial mucin (sialomucin) associated disease can be excluded. Although papular mucinosis (lichen myxoedematosus) can occur locally rather than in a generalized form, and the pattern of mucin distribution may be diffuse or focal, involving the upper and mid reticular dermis. However, this disease belongs to one kind of degenerative-inflammatory mucinosis and affects adults, most commonly seen in patients 30 to 50 years old. No congenital case was reported in the literature. Reticular erythematous mucinosis is a persistent, photoaggrevated, erythematous, reticular or plaque-like eruption in the middle of the back or chest and occurs most often in middle-aged women. Clinically, our case is different from this disease. Acral persistent papular mucinosis is characterized by multiple ivory- or flesh-colored papules, which only occur on the extensor surface of hands and forearm. All cases reported are adult and predominate in female patients. Both location and age are not compatible with this case. Self-healing juvenile cutaneous mucinosis only occurs in children and is characterized by acute eruption of multiple papules which aggregate in linear, infiltrated plaques, giving a corrugated appearance to the skin and followed by spontaneous resolution within a few months. It may be accompanied with some associated symptoms, such as fever, arthralgia, muscle tenderness and weakness. Obviously, clinical course is different from this case. Cutaneous focal mucinosis is a reactive lesion and occurs in adulthood. It is characterized by a solitary white or flesh-colored papule or nodule. It can appear anywhere over the body, except the joints of hands and feet. No congenital case was reported in the literature. Histopathologically, mucinous changes are not confined to the hair follicle in our case (Fig. 2, 3). So follicular mucinosis was ruled out.

Cutaneous myxoma (angiomyxoma) is an aquired benign neoplasm. It can be solitary or multiple. However, histopathologically, it is a moderately well-demarcated and multilobulated lesion characterized by a mucinous matrix involving the dermis and subcutis. Since the result of histopathological examination is different from this disease, cutaneous myxoma was ruled out. The most important differential diagnosis for our case is cutaneous mucinosis of infancy (CMI). CMI is also a rare condition of unknown etiology, characterized by small papular mucin deposits which are either symmetrical and densely grouped on the arms or linear and localized, and are congenital or appear in early infancy. In fact, 1993 Redondo Bellon et al., first described MN, they assigned it to the spectrum of CMI. But recently some authors believed that mucinous lesions with naevoid features, namely their unilateral and/or linear or dermatomal pattern better satisfy the criteria of MN. In addition, adolescent or adult onset cases cannot be included in an infantile group. Therefore, in the newest classification of cutaneous mucinosis made by Rongioletti and Rebora, MN and CMI belong to different subtypes of primary cutaneous mucinosis. CMI is a kind of degenerative-inflammatory mucinosis. And it has recently been suggested that this condition might represent a pediatric form of lichen myxoedematosus. On the other hand, MN is hamartomatous mucinosis. Given the striking grouped nodules with band-like distribution and congenital onset, other kinds of CM were ruled out. We diagnosed this case as mucinous nevus.

Mucinous nevus is a very rare entity. MN was first described by Redondo Bellon et al. in 1993 and can be classified as both a cutaneous mucinosis (CM) and a connective tissue nevus (CTN) due to they have overlapping features. Cutaneous mucinosis is a heterogeneous group of diseases. According to the most recent comprehensive classification of the histopathologic
and clinical characterization for the cutaneous mucinosis updated and published by Rongioletti and Rebora\(^2\) in 2001, CM can be divided into two groups: I, distinctive (primary) CM and II, diseases associated with histopathologic mucin deposition (secondary mucinosis). The former is further divided into degenerative-inflammatory mucinosis and neoplastic-hamartomatous mucinosis. MN belongs to the neoplastic-hamartomatous mucinosis of primary mucinosis. CTN is a hamartomatous condition consisting of various components of the dermal connective tissue. The predominant element of the extracellular connective tissue within an individual nevus can be collagen, elastic fibers, or proteoglycan (mucin). CTN of the proteoglycan type includes an inherited form (mucopolysaccharidosis), while the acquired type represents some forms of CM: cutaneous focal mucinosis and lichen myxedematosus. Due to the overlapping features, MN was thought to be a variant of either CTN of proteoglycan type or primary distinctive CM. In our opinion, we prefer the former because we can know that it is a hamartomatous condition and consists of abundant proteoglycan (mucin) in the dermis just from the name. To our knowledge, only 10 cases identified as MN in English literature\(^1,4,6-11\) (Table I.) and here we identified the first case in Taiwan.

We review 10 cases reported in English literature (Table I.) and find they share certain clinical features. They appear to consist of multiple brownish to flesh-colored papules or plaques with a striking unilateral and linear or zosteriform distribution. The lesions develop at birth, adolescence or early adulthood. Both size and number may slowly increase with time. Coalescence was reported in some cases. The lower part of the trunk is the most commonly affected site. Most cases appear as teenager. Interestingly it seems that males predominate over females. Of the 10 reported patients, 9 are male. It is asymptomatic, although repeated minor trauma may produce secondary changes within the lesions.\(^6\) Fortunately, so far, no associated anomalies or systemic illnesses have been reported.

The hallmark of histopathological examination is prominent interstitial mucin deposit in the dermis. Although papillary and superficial dermis was the main site of mucin deposition in most reported cases, it is reasonable that mucin can deposit in any portion of dermis in MN. It is just like other kinds of CTN, in which the location of collagen or elastic tissue is not limited to certain portion of dermis because they are hamartomatous conditions. The reasonable explanation of why mucin deposited in papillary and superficial dermis in most reported cases is that the more superficial the mucin deposit, the more obvious the clinical presentation is. We believe that it is a result of underdiagnosis and mucin can deposit anywhere in the dermis of MN. Other findings may include acanthosis, often with thin, elongated rete ridges, a paucity of collagen and elastic fibers in the papillary dermis, slightly increased fibroblasts, and a sparse perivascular lymphocytic infiltrate. As mucin staining gives a positive reaction with alcian blue at pH 2.5, but is negative at pH 0.5, and positive reaction with colloidal iron, the mucin in the lesion is thought to be composed of hyaluronic acid.\(^6\) The origin of the mucin deposited in the lesion remains unclear. Because the number of fibroblast increases in most reported cases and lesions developed early in life, even at birth, without evidence of trauma or a pre-existing pathological changes at the site of the lesion and the size and number of lesions become larger during long term following up, we propose that the mucin is overproduced by these abnormal fibroblasts.

Since the epidermal changes are not a constant feature of MN, some authors have suggested that they should be further divided into two different subtypes. MN without epidermal nevus-like changes should be included under CTN of the proteoglycan type while MN with epidermal changes should be included in the combined epidermal-CTN of proteoglycan type. After reviewing all reported cases, we find that combined epidermal-CTN of proteoglycan type is predominant. Of the 10 reported patients, 8 belong to this subtype.
### Table 1. Mucinous Nevus in the English-language Literature

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Lesion pattern</th>
<th>Distribution Location</th>
<th>Onset Age</th>
<th>A/R/S</th>
<th>Increased fibroblast deposition</th>
<th>Superficial dermis</th>
<th>Papillary dermis</th>
<th>Epidermal change*</th>
<th>Reference No.</th>
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<tr>
<td>1</td>
<td>papules</td>
<td>Zosteriform arrangement</td>
<td>Interscapular area</td>
<td>Congenital</td>
<td>16 y/o/F</td>
<td>+</td>
<td>Superficial dermis</td>
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<td>2</td>
<td>papules</td>
<td>Zosteriform arrangement</td>
<td>Lower back</td>
<td>19 y/o</td>
<td>23y/o/M</td>
<td>+</td>
<td>Superficial dermis</td>
<td>+</td>
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<td>plaque</td>
<td>Zosteriform arrangement</td>
<td>Lower back</td>
<td>22 y/o</td>
<td>27y/o/M</td>
<td>+</td>
<td>Papillary dermis</td>
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<td>4</td>
<td>Brown nodules</td>
<td>Zosteriform arrangement</td>
<td>Right abdomen</td>
<td>Adolescence</td>
<td>61y/o/M</td>
<td>+</td>
<td>Papillary dermis</td>
<td>+</td>
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<td>papules</td>
<td>Zosteriform arrangement</td>
<td>R’t lower back</td>
<td>10 y/o</td>
<td>17y/o/M</td>
<td>+</td>
<td>Papillary dermis</td>
<td>+</td>
<td>7</td>
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<td>6</td>
<td>plaque plagues</td>
<td>Zosteriform arrangement</td>
<td>L’t lower back</td>
<td>Congenital</td>
<td>14y/Korean/M</td>
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<td>Papillary dermis</td>
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<td>papules</td>
<td>Nevoid arrangement</td>
<td>L’t buttock</td>
<td>10 y/o</td>
<td>24y/o/M</td>
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<td>+</td>
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<tr>
<td>8</td>
<td>plaque plagues</td>
<td>Linear arrangement</td>
<td>Lower back</td>
<td>?</td>
<td>30y/o/M</td>
<td>-</td>
<td>Papillary dermis</td>
<td>+</td>
<td>9</td>
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<tr>
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<td>papules</td>
<td>Linear arrangement</td>
<td>R’t upper chest</td>
<td>13y/0</td>
<td>15y/0/M</td>
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<td>Papillary dermis</td>
<td>+</td>
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<tr>
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<td>papules</td>
<td>Linear arrangement</td>
<td>Back and abdomen</td>
<td>Congenital</td>
<td>14M/Asian/M</td>
<td>+</td>
<td>Papillary dermis</td>
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</table>

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Similar to other hamartomas, MN does not need to be treated, but it can be removed if it is symptomatic or cosmetically disfiguring. Surgical excision without recurrence was reported.

REFERENCES