Combined epidermal-connective tissue nevus of proteoglycan (a type of mucinous nevus): a case report and literature review

Mucinous nevus is a rare entity with only 11 cases reported previously. It may be divided into two histopathologic types: connective tissue nevus of the proteoglycan (CTNP) and combined epidermal-CTNP. We describe a boy with asymptomatic grouped brown papuloplaques on the lower back since birth. A diagnosis of mucinous nevus of the combined epidermal-CTNP type was made after a biopsy. We vaporized two lesions with carbon dioxide laser, and the wounds healed satisfactorily. We present a literature review indicating a striking preponderance of male patients (M:F = 5:1) for mucinous nevus. In half of the cases, mucinous nevus did not appear until childhood, adolescence or early adulthood. The predominantly affected site was the trunk. Half of the cases can be assigned to the CTNP type and the other half to the combined epidermal-CTNP type. We propose that carbon dioxide laser vaporization may be a treatment option for mucinous nevus of the combined epidermal-CTNP type with multiple lesions but not for the CTNP type.


Mucinous nevus is a rare entity first described by Redondo Bellón et al. in 1993. To the best of our knowledge, there have been only 11 cases of mucinous nevus reported previously. Mucinous nevus is divided into two histopathologic types: connective tissue nevus of the proteoglycan (CTNP) type and combined epidermal-CTNP type. In this study, we describe a case of combined epidermal-CTNP that clinically resembled epidermal nevus and present a review of the literature.

Case report
A 17-year-old boy visited our outpatient clinic in January 2005 and presented with asymptomatic grouped brown papules and coalescent plaques on his middle lower back since birth (Fig. 1). Under the clinical impression of epidermal nevus, a skin biopsy was performed. Microscopically, the specimens showed marked invaginating hyperkeratosis, elongated rete ridges, pigmented basal cell layer and loosely separated collagen fibers in papillary dermis suggesting mucinosis or edema. Some fatty tissue components were also present in the upper dermis (Fig. 2A). After processing with alcian blue at pH 2.5, positively stained amorphous materials were found in the papillary dermis (Fig. 2B), but the staining was removed with hyaluronidase digestion, proving that these amorphous deposits were mucin. Focal disruption of elastic network in the papillary dermis was shown by elastic stain (Fig. 2C). Considering the clinical manifestations and pathologic findings, a diagnosis of mucinous nevus of combined epidermal-CTNP type was made.

We vaporized two lesions with a carbon dioxide laser, and the wounds healed well with no signs of recurrence after a follow up for 6 months. Because of the lack of cosmetic significance with the lesions being confined to low back, the patient declined further treatment.

Discussion
The previously reported cases of mucinous nevus and the present one are summarized in Table 1.1–9 We observed a striking preponderance of male patients (M : F = 5 : 1), but the reason is not known. In half of the reported cases, mucinous nevus was not present at birth and developed in childhood, adolescence or early adulthood.2,3,5–7 Except for one case,9 the site of involvement was the trunk. The typical clinical presentation of mucinous nevus is grouped brownish papules and confluent plaques, usually arranged in a unilateral, linear, zosteriform or grouped distribution. The nevoid feature of mucinous nevus is clinically indistinguishable from epidermal nevus, other connective tissue nevus or nevus lipomatosus superficialis.

Redondo Bello´n et al. included mucinous nevus among the spectrum of cutaneous mucinosis of infancy.1 However, in contrast to cutaneous mucinosis of infancy with small papular lesions, the loci of mucinous nevus are in a nevoid distribution and composed of papules and confluent plaques. Rongioletti and Rebora examined two previously reported cases of cutaneous mucinosis of infancy and reclassified them as mucinous nevus.8,9

Connective tissue nevus is a hamartoma composed of excessive amounts of one or several components of the dermis, such as collagen and elastin, and is specified as collagenoma and elastoma, respectively. It may appear at birth or in early childhood.10 Brakman et al. regarded mucinous nevus as a distinct type of connective tissue nevus of proteoglycan (mucin).6,7

Connective tissue nevus such as collagenoma may be associated with other congenital anomalies, for example osteopoikilosis (in Buschke-Ollendorf syndrome),
tuberous sclerosis and focal dermal hypoplasia (i.e. Goltz syndrome). However, our literature review did not reveal an association between mucinous nevus and congenital anomalies.

The primary histopathologic feature of mucinous nevus is diffuse mucin deposition in the superficial dermis, usually in a band-like distribution that accords with its clinical papuloplaque lesions. The mucin is judged to be hyaluronic acid because of having positive reactivity with alcian blue at pH 2.5 but negative reactivity at pH 0.5 and removal of the staining by hyaluronidase. The origin of excessive mucin remains unclear. Although mucin is synthesized by fibroblasts, only a slight increase of activated fibroblasts has been illustrated in previously reported cases. Some authors therefore suggested that the upregulation of fibroblasts is responsible for the overproduction of mucin. Disruption of collagen and elastic fiber network in the mucinous area is often observed as well.

Mucin deposition in the papillary dermis is also seen in cutaneous mucinosis of infancy but is often very superficial and appears to be ‘hugged’ by the epidermis that conforms with its clinical smaller papular lesions.

Mucin deposition in the papillary dermis is also seen in cutaneous mucinosis of infancy but is often very superficial and appears to be ‘hugged’ by the epidermis that conforms with its clinical smaller papular lesions. The epidermis may either be normal or show hyperkeratosis and acanthosis with elongation of rete ridges indicating epidermal nevus. Rongioletti and Rebora classified the former type of mucinous nevus as CTNP and the latter type as combined epidermal-CTNP. After reviewing the histopathologic changes of the reported cases of mucinous nevus, we found that half of them, including ours, can be ascribed to the CTNP type and the other half to the combined epidermal-CTNP type.

Mucinous nevus of both the CTNP and the combined epidermal-CTNP types may be treated with surgical excision. However, excision is often not feasible for those with multiple discrete lesions. The feasibility of a treatment may also vary with the pathologic type of the mucinous nevus. Yokogawa et al. treated a mucinous nevus of the CTNP type with scalpel dermabrasion, but scars were noted 1 year later. In the present case of the combined epidermal-CTNP type, we treated a couple of exophytic lesions with carbon dioxide laser vaporization and achieved acceptable cosmetic results. However, laser vaporization may not be appropriate for treating mucinous nevus of the CTNP type where the epidermis is normal and might produce scars.

In conclusion, we found a striking preponderance of male patients (M : F = 5 : 1) for mucinous nevus. In half of the cases, mucinous nevus did not appear at birth but developed in childhood, adolescence or early adulthood. The affected site was predominantly the trunk. Half of the cases can be classified as the CTNP type and the other half as the combined epidermal-

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**Table 1. Reported cases of mucinous nevus**

<table>
<thead>
<tr>
<th>Author</th>
<th>Gender</th>
<th>Age at onset</th>
<th>Location</th>
<th>Configuration</th>
<th>Classification</th>
<th>Epidermal pathology</th>
<th>Treatment</th>
<th>Follow up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Redondo Bello´ n et al.</td>
<td>Female</td>
<td>Birth</td>
<td>Intercapilar</td>
<td>Grouped</td>
<td>Combined epidermal-CTNP</td>
<td>Epidermal nevus like</td>
<td>Excision</td>
<td>No recurrence 8 months later</td>
</tr>
<tr>
<td>Braakman et al.</td>
<td>Male</td>
<td>19 years</td>
<td>Back</td>
<td>Linear</td>
<td>Combined epidermal-CTNP</td>
<td>Epidermal nevus like</td>
<td>Excision</td>
<td>No recurrence 4 months later</td>
</tr>
<tr>
<td>Rongioletti and Rebora</td>
<td>Male</td>
<td>22 years</td>
<td>Lower back</td>
<td>Linear</td>
<td>Normal</td>
<td>Epidermal nevus like</td>
<td>Excision</td>
<td>No recurrence 6 months later</td>
</tr>
<tr>
<td>Carpezi et al.</td>
<td>Male</td>
<td>22 years</td>
<td>Fingers</td>
<td>Linear</td>
<td>Normal</td>
<td>Epidermal nevus like</td>
<td>Excision</td>
<td>No recurrence 6 months later</td>
</tr>
<tr>
<td>Carpezi et al.</td>
<td>Male</td>
<td>Birth</td>
<td>Adolescence</td>
<td>Grouped</td>
<td>Combined epidermal-CTNP</td>
<td>Epidermal nevus like</td>
<td>Excision</td>
<td>No recurrence 4 months later</td>
</tr>
<tr>
<td>Chang et al.</td>
<td>Male</td>
<td>10 years</td>
<td>Lower back</td>
<td>Grouped</td>
<td>Normal</td>
<td>Epidermal nevus like</td>
<td>Excision</td>
<td>No recurrence 3 months later</td>
</tr>
<tr>
<td>Lim et al.</td>
<td>Male</td>
<td>Not stated</td>
<td>Lower back</td>
<td>Linear</td>
<td>Normal</td>
<td>Epidermal nevus like</td>
<td>Excision</td>
<td>No recurrence 6 months later</td>
</tr>
<tr>
<td>Yokogawa et al.</td>
<td>Male</td>
<td>13 years</td>
<td>Right upper chest</td>
<td>Linear</td>
<td>Normal</td>
<td>Epidermal nevus like</td>
<td>Excision</td>
<td>No recurrence 6 months later</td>
</tr>
<tr>
<td>Chi et al. (this study)</td>
<td>Male</td>
<td>Birth</td>
<td>Lower back</td>
<td>Linear</td>
<td>Normal</td>
<td>Epidermal nevus like</td>
<td>Two lesions treated with carbon dioxide laser</td>
<td>No recurrence 6 months later</td>
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</tbody>
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CTNP, connective tissue nevus of the proteoglycan type.

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CTNP type. The applicability of a treatment may vary with the pathologic type of the lesion. Carbon dioxide laser vaporization may be a treatment option for mucinous nevus of the combined epidermal-CTNP type with multiple exophytic and verrucous lesions but not for the CTNP type.

References
