Those who support the connective tissue origin will further divide mucinous nevi into 2 histopathologic types: a connective tissue nevus of the proteoglycan (CTNP) type and combined epidermal-CTNP type.1 In CTNP, the epidermis is normal, while in epidermal-CTNP, the epidermis shows hyperkeratosis and acanthosis with elongation of the rete ridges consistent with an epidermal nevus.3 Regardless of classification, the origin of increased mucin is unclear. Although mucin is synthesized by fibroblasts, previous studies have shown only a slight increase in activated fibroblasts. Thus, some have postulated that the upregulation of fibroblasts is responsible for the increased mucin.3

Clinically, these lesions can be hard to distinguish from connective tissue nevi such as collagenomas or elastomas. Histologic examination is essential to exclude these entities, which can be associated with congenital abnormalities. The microscopic differential diagnosis is narrow and includes focal cutaneous mucinosis and lichen myxedematosus, both of which have distinct clinical presentations. Lesions of focal cutaneous mucinosis are usually solitary papules and can be found anywhere on the body. Lichen myxedematosus presents with a slow onset of asymptomatic or slightly pruritic papules, which can be generalized or localized. Generalized subtypes of lichen myxedematosus have an associated monoclonal gammopathy, while discrete papular lichen myxedematosus, a variant of the localized subtype, has been associated with human immunodeficiency virus disease.6 Other histologic mimics, like self-healing papular mucinosis and acral persistent papular mucinosis, are easily excluded with clinical information. In our case, clinical correlation confirmed the rare diagnosis of mucinous nevus.

Treatment for a mucinous nevus is not required owing to its benign nature. Surgical intervention may remove the lesion, but scarring will result.

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Poland Syndrome Coexisting With Blaschkolinear Congenital Melanocytic Nevi

Poland syndrome is an uncommon congenital anomaly characterized by unilateral chest wall and upper extremity hypoplasia.1 It is theorized to be due to subclavian artery hypoplasia during embryogenesis.1 There have been only a few isolated reports of Poland syndrome with congenital dermatoses.2-4 Adding to this association list, we report a case of Poland syndrome with multiple congenital melanocytic nevi (CMN). Additionally, the nevi are in a Blaschkolinear arrangement, which is also rare; we know of only 2 such cases reported previously.5,6

Report of a Case | An otherwise healthy boy in his teens presented for asymptomatic hyperpigmented birthmarks on his left leg that had grown proportionately to his leg’s growth. His medical history was significant for Poland syndrome, for which he had undergone multiple left-hand reconstruction surgeries. His family history was noncontributory. Physical examination revealed oligodactyly of the left hand, underdeveloped left chest wall (Figure 1A), and multiple, evenly pigmented, bluish-black thin papules and plaques on the left posterior thigh, calf, and ankle in a Blaschkolinear distribution (Figure 1B). A

Figure 1. Clinical Images of Patient With Poland Syndrome and Blaschkolinear Congenital Melanocytic Nevi

A. Hypoplasia of the left chest wall and oligodactyly of the left hand. The 2 digits of left hand seen in the photograph were reconstructed from his toes. B. Blaschkolinear distribution of bluish-black thin papules and plaques on his left leg.
skin biopsy specimen from the left posterior thigh showed characteristics consistent with CMN (Figure 2).

Discussion | Poland syndrome, named after the British surgeon Alfred Poland who first described it in 1841, is an uncommon, sporadic, and very rarely inherited birth defect characterized by unilateral chest wall hypoplasia (often right-sided) and ipsilateral hand deformity (most often syn-brachydactyly and less often oligodactyly).1 Poland syndrome affects boys 2 to 3 times as often as girls, with an estimated incidence of 1 in 10,000 to 100,000 live births. Its exact cause remains unclear, but a prevailing theory is hypoplasia of the subclavian artery during the critical sixth week of gestation, which leads to a range of musculoskeletal malformations.1

Poland syndrome has been associated with hematopoietic malignancies such as leukemia and non-Hodgkin lymphoma, as well as other syndromes, including Möbius syndrome (characterized by congenital bilateral facial paralysis with inability to abduct the eyes) and Klippel-Feil syndrome (characterized by congenital fusion of any 2 of the 7 cervical vertebrae).3 In addition to these known associations, a literature review revealed a few isolated cases of Poland syndrome with various congenital dermatologic findings, including recessive X-linked ichthyosis, congenital hemangioma, and café-au-lait spots.3-4 To our knowledge, the case is the first reported case of Poland syndrome associated with multiple CMN.

A CMN is a benign, clonal proliferation of melanocytes in the epidermal, dermal, or subcutaneous tissue that is present at or shortly after birth, with an estimated prevalence of 0.5% to 31.7%, depending on the study. They are usually solitary, but 3% are multiple, occasionally arranged in a cluster and rarely in a linear distribution. Indeed, only 2 cases of Blaschko linear CMN have been reported thus far.5,6 Our patient would represent the third such case, and it occurs in a patient with Poland syndrome.

The definite risk of developing melanoma in Blaschko linear CMN remains unclear because of its rarity. Our patient had no clinical sign of malignant transformation by age 11 years, as in the 2 previously reported cases at ages 15 and 28 years. However, long-term close clinical monitoring might be warranted, as in giant CMN.

In summary, we report herein a rare case of Blaschko linear CMN in a teenager with Poland syndrome. The association between the 2 conditions seems to be coincidental, as in previously reported cases of Poland syndrome with other congenital dermatologic findings.