

Linear Unilateral Basal Cell Nevus: A Benign Follicular Hamartoma Simulating Multiple Basal Cell Carcinomas

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We report a case of linear unilateral basal cell nevus (LBCN) occurring on the left lateral neck and left posterior shoulder of a 23-year-old woman. LBCN is a rare benign follicular hamartoma that must be distinguished from the more aggressive unilateral and segmental variant of nevoid basal cell carcinoma syndrome (NBCCS) and the linear variant of BCC.

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Linear unilateral basal cell nevus (LBCN) is a rare benign condition that can histologically mimic basal cell carcinoma (BCC). We present the case of a woman with LBCN and discuss the relevant literature, including ways to differentiate LBCN from variants of nevoid BCC syndrome (NBCCS) and BCC.

Case Report

A 23-year-old white woman presented with a linear array of 1- to 5-mm pearly, dome-shaped, nevoid papules on the left lateral neck that had been present without significant change since adolescence. Several of the papules were studded with dark pigment (Figure 1). She also had a linear band of hypopigmented macules and papules extending from the left posterior shoulder down the left arm (Figure 2). Palmar and plantar pitting and

dysmorphic facial features were absent. Hair, teeth, and nails were unaffected. Prior chest and dental radiographs were within reference range. There was no personal or family history of skin cancer or other related abnormalities.

Examination of 10 different biopsy specimens from the neck and shoulder demonstrated nodular masses and strands of basaloid cells with areas of palisading peripheral nuclei and retraction from the surrounding stroma. Approximately 2 years after presentation, the lesions had not shown any progression and further treatment had not been pursued.

Comment

In 1952, Carney¹ first reported a case of LBCN with comedones in a 69-year-old white man. The hyperpigmented linear lesions were noted at birth and involved the left scalp, cheek, chest, abdomen, arm, and leg. Comedones and smooth globular papules with a pearly appearance were noted. Biopsy results were consistent with basal cell epithelioma.

LBCN is a rare entity with few case reports since Carney's¹ initial publication. Lesions commonly present at birth,¹⁻⁶ but they may appear during early adulthood.⁷⁻⁹ The lesions are distributed unilaterally on the face, trunk, and extremities, and appear to follow Blaschko lines.^{4,10} In addition to smooth pearly papules, other reported cutaneous findings included epidermoid cysts, verrucous papules, atrophic areas, and comedones.¹⁻⁷ The multiplicity of lesions noted clinically emphasizes the hamartomatous nature of LBCN.¹⁰ Biopsy specimens of the lesions have been indistinguishable from various histologic types of BCC but without clinical demonstration of aggressive behavior.^{3,5,7,9} There is considerable clinical and histologic overlap with basaloid follicular hamartoma (BFH) and LBCN.¹⁰ Three forms of BFH have been described and one can present in a

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Figure 1. Pearly, flesh-colored, hyperpigmented papules in a linear arrangement on the left lateral neck.



Figure 2. Linear band of hypopigmented macules and papules on the left posterior shoulder and arm.

linear and unilateral fashion.^{6,9} LBCN can be synonymous with the linear and unilateral presentation of BFH.⁹

Reports of associated anomalies including scoliosis, anodontia, osteoma cutis, and abnormal bone mineralization raise the possibility that LBCN may be considered a variant of epidermal nevus with additional features analogous to the epidermal nevus syndrome.^{1,7,8,10,11} However, the paucity of case reports and lack of a unifying theme do not allow firm conclusions to be drawn at this time. No familial pattern has been described for LBCN. In contrast, the NBCCS is inherited in an autosomal dominant fashion and results from mutations in the patched gene.¹² This syndrome is associated with numerous anomalies, including multiple aggressive BCCs presenting at an early age, jaw cysts, skeletal anomalies, ectopic calcification, and palmar and plantar pitting.¹³ Unilateral or segmental manifestations of NBCCS have been previously reported.¹³⁻¹⁵ In addition to multiple unilateral BCCs, these patients variably expressed other anomalies commonly seen in NBCCS, including jaw cysts, palmar pits, skeletal deformities, facial dysmorphism, and calcification of the falx cerebri.¹³⁻¹⁵ Because of the lack of family history in these patients, it is assumed that these abnormalities were derived from new postzygotic somatic mutations.^{10,12} The benign behavior of the basaloid proliferations and absence of other cutaneous findings such as atrophic areas and comedones can help distinguish the lesions

of LBCN from NBCCS. Furthermore, the few anomalies associated with LBCN are not commonly observed in patients with NBCCS.

BCC in sun-damaged skin may develop in a linear pattern along relaxed skin tension lines.^{16,17} Linear BCC is more clinically aggressive than LBCN. The patient's history also can help differentiate linear BCC from LBCN. The presence of the lesions from an early age without significant progression argues for the latter diagnosis. Mohs micrographic surgery is the treatment of choice for linear BCC.¹⁶ To our knowledge, invasive behavior in the lesions of LBCN has not been documented. Therefore, aggressive treatment of these lesions does not appear to be warranted. However, because of reports of progressive BCC developing in various types of epidermal nevi, it is prudent to provide close follow-up in patients with LBCN.¹⁸

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