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Authors

Atmatzidis, Douglas H
Zouzias, Christos
Rogachefsky, Arlene
et al.

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Rippled-pattern basal cell carcinoma: a unique characteristic of an otherwise common diagnosis

Douglas H Atmatzidis MD, Christos Zouzias MD MBA, Arlene Rogachefsky MD, Kristyna Lee MD MPH

Affiliations: Affiliated Dermatologists & Dermatologic Surgeons, Professional Association, Morristown, New Jersey, USA

Corresponding Author: Douglas H Atmatzidis MD, Affiliated Dermatologists & Dermatologic Surgeons PA, 182 South Street, Suite 1, Morristown, NJ 07960, Tel: 973-267-0300, Email: douglas.atmatzidis@gmail.com

Abstract

Herein we present a 48-year-old man who was diagnosed with a nodular basal cell carcinoma (BCC) of the left chin that was revealed on Mohs frozen sections to be a rippled-pattern BCC with invasion into muscle and necessitated interdisciplinary care for concern for a concurrent salivary gland carcinoma. Rippled-pattern BCC has been documented in fewer than ten cases, though this may also be attributed to confusion regarding nomenclature. Microscopically, rippled-pattern BCC exhibits distinctive features that differentiates it from other patterns. The tumor cells form interconnected networks or cords with intervening mucinous spaces within a fibrotic stroma resulting in a rippled appearance. As they are generally considered nodular BCCs, management of rippled-pattern BCCs can involve surgical excision, Mohs micrographic surgery, or other appropriate treatment modalities based on the tumor's location, size, and degree of invasion.`

Keywords: basal, Mohs, rippled

Introduction

Rippled-pattern basal cell carcinoma is a poorly documented pattern of basal cell carcinoma (BCC) that presents unique histopathological features. Although rippled-pattern BCC is not considered an aggressive feature and has a similar prognosis to nodular BCC [1], we present a healthy man with a rippled-pattern BCC that demonstrated invasive behavior and required interdisciplinary management with extensive surgical repair.

Case Synopsis

A 48-year-old man presented with a five-year history of a 0.5cm discrete pearly pink papule on his left chin in a background erythematous patch across the rest of his chin (**Figure 1A**). A biopsy revealed BCC, nodular type (**Figure 1B**). During three stages of Mohs micrographic surgery (MMS), frozen sections revealed evidence of nodular BCC, but in some areas tumor cells formed reticulated and cystic patterns surrounded by pseudo-tubular cord-like structures and mucinous clefts (**Figure 2A-C**). No perineural invasion was seen. The resultant defect extended to peripheral and deep margins, including muscle (**Figure 1C**). Given the unique histological findings and degree of invasion, there was concern for a concurrent salivary gland carcinoma. Thus, Mohs surgery was halted and the patient was referred to the head and neck surgery department for further treatment. The slides were sent for permanent sections and consultation with a dermatopathologist was consistent with a diagnosis of rippled-pattern BCC. No salivary gland carcinoma was identified. The patient underwent wide local excision by a head and neck surgeon revealing additional tumor at peripheral margins (final defect 4.3cm×3.4 cm) and local tissue reconstruction was accomplished with a cervical flap.

Case Discussion

Basal cell carcinoma is the most common skin cancer worldwide, typically occurring in sun-exposed areas, particularly the face, and often presenting as a slowly

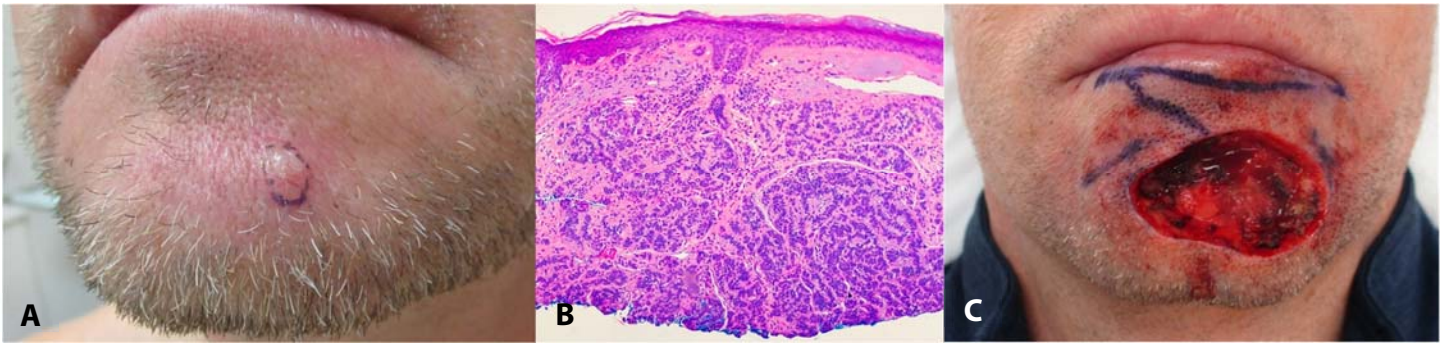

Figure 1. ***A)*** *Tumor at the time of biopsy. Five-millimeter papule on the left chin circled in gentian violet marker. Note the subtle erythematous patch extending over much of the chin.* ***B)*** *H&E histopathology of biopsy, 40×. In the dermis is a circumscribed proliferation of basaloid lobules and rounded cords in a fibromyxoid stroma with clefting between tumor and stroma.* ***C)*** *Post-Mohs Defect. The defect extended into the mentalis and inferior portion of the orbicularis oris.*

growing, firm, pink or flesh-colored papule or nodule [2,3]. Well-known subtypes include superficial, nodular, micronodular, and infiltrative. We seek to explore the less-frequently encountered rippled-pattern histology.

Despite how common BCCs are, to our knowledge, the rippled-pattern BCC has been documented in fewer than ten cases [1]. Criteria proposed for the histopathological characteristics of the rippled-pattern BCC include the following: a nodular type BCC, frequent intervention by mucinous spaces between the epithelial cords, and lack of derivation from adnexal structures [1].

However, confusion may exist regarding terminology, as the descriptors neuroid-type and schwannoid BCC can be found in the pathology literature, which describe the same phenomenon [4]. Microscopically, rippled-pattern BCC exhibits distinctive features that differentiate it from other patterns. The tumor cells form interconnected networks or cords with intervening mucinous spaces within a fibrotic stroma resulting in a rippled appearance.

The histopathological similarities between rippled-pattern BCC and other cutaneous neoplasms may pose challenges in differential diagnosis. It should be distinguished from other tumors, particularly rippled-pattern sebaceoma and trichoblastoma [1,4]. Compared to sebaceomas, rippled-pattern BCCs can be differentiated by negative staining of the tumor cells for epithelial membrane antigen [1]. Although androgen receptor (AR) displays diffuse expression

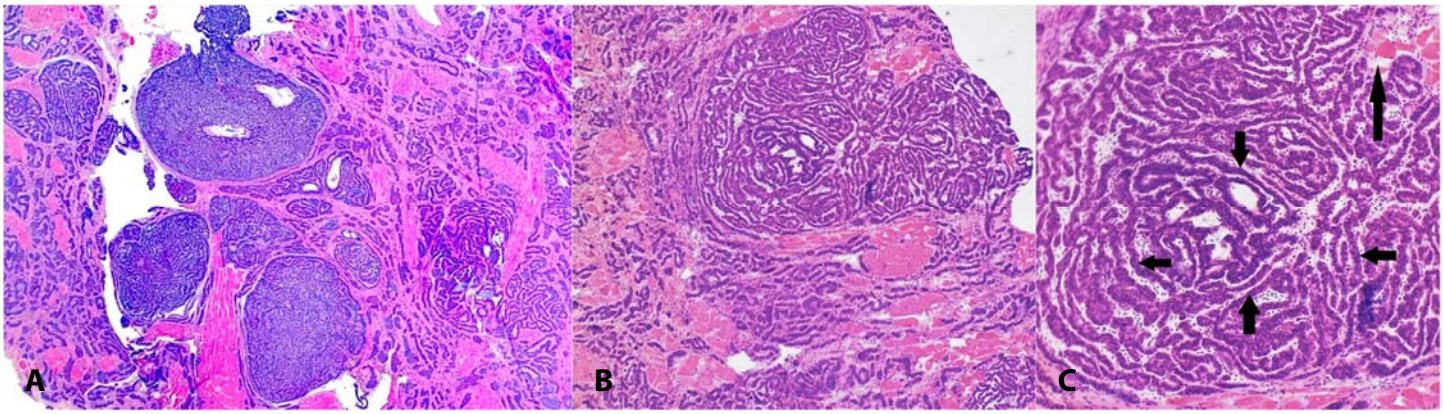

Figure 2. *H&E histopathology.* ***A)*** *Mohs permanent section, 40×. Tumor in dermis and invading skeletal muscle composed of basophilic keratinocytes that form loose and compact interconnected networks or cords with intervening mucinous spaces within a fibrotic stroma resulting in a "rippled" appearance.* ***B)*** *Mohs frozen section, 100×. Higher magnification image showing details of the tumor nodule with "rippled" appearance.* ***C)*** *Mohs frozen section, 200×. Short black arrows pointing to the alternating bands of epithelial cords of spindle-shaped basaloid cells and mucinous spaces arranged in parallel rows. Long black arrow denoting clefting of the tumor from the surrounding stroma characteristic of nodular basal cell carcinoma.*

in sebaceous tumors and focal expression in BCC, Ber-EP4 is typically negative in sebaceoma and positive in BCC [5]. Although adipophilin and perilipin have been shown to stain strongly in sebaceomas and are restricted to sebocytes in BCC with sebaceous differentiation, nonspecific staining may be an obstacle [5].

Trichoblastoma stains positive for stromal CD10 and CD34 as well as pleckstrin homology-like domain family A member one (PHLDA1), a follicular stem cell marker, whereas BCC typically does not [6]. Cytokeratin 20 (CK20) can be used in combination with AR, with positive tumor AR staining and absent CK20 staining supporting BCC, whereas $CK20^-$ positive colonizing Merkel cells and lack of AR support a diagnosis of trichoblastoma [5]. Although AR, CK20, and PHLDA1 may be useful in distinguishing BCC from benign hair-germ tumors, there appears to be no single marker completely sensitive or specific for this distinction [5].

Biswas et al. described the various additional tumors that have rippled-pattern cases documented, including dermatofibroma, dermatofibrosarcoma protuberans, schwannomas, leiomyomas, melanocytic nevi, melanomas, pleiomorphic adenoma, and intranodal palisaded myofibroblastoma [4]. Routine histologic analysis and immunohistochemistry, if needed, should aid in differentiation from these other tumors [4]. As they are generally considered nodular BCCs, management of rippled-pattern BCCs can involve surgical excision, MMS, or other appropriate treatment modalities based on the tumor's location, size, and degree of invasion [1].

Rippled-pattern BCC typically has a favorable prognosis, but as is demonstrated in our case, early diagnosis is critical for better outcomes. The subtle surrounding erythematous patch around the initially-biopsied papule demonstrated in **Figure 1A** hinted at the eventual peripheral extent of the tumor, but the deep invasion into the mentalis and orbicularis oris muscle was unexpected.

Conclusion

Rippled-pattern BCC is a distinct histological feature of some nodular BCCs. Despite its rarity, dermatologists and Mohs surgeons should be aware of this appearance to ensure accurate diagnosis and appropriate management. Further research and clinical studies are necessary to enhance our understanding of rippled-pattern BCC and its optimal treatment.

Potential conflicts of interest

The authors declare no conflicts of interest.

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