

A Rare Collision in Dermatopathology: Basal Cell Carcinoma and Atypical Fibroxanthoma

To the Editor:

We present the second report of a rare collision tumor comprised of a basal cell carcinoma (BCC) and an atypical fibroxanthoma (AFX) in a 64-year-old white man, who presented for evaluation of a bleeding growth on his right temple. The lesion had been present for several years but was becoming larger and increasingly pruritic over the past 3 months. On physical examination, a 3.5-cm pink, pearly irregularly shaped plaque was noted over the right temple, with light brown crust along the edges. A hyperpigmented focus was noted at the inferior aspect of the plaque with overlying erosions (Fig. 1).

Histologic examination revealed a mass with 2 different histologic patterns (Fig. 2). One part of the tumor exhibited features consistent with a classic BCC, comprised of islands and nests of basaloid cells, originating from the basal layer of the epidermis and extending into the dermis, with peripheral palisading and focal epidermal-stromal clefting, surrounded by a mucinous stroma (Fig. 3). Adjacent to and intermixed with the first tumor was an infiltrating spindle cell proliferation in a haphazard arrangement that abutted the epidermis but did not appear to be connected to it (Figs. 4, 5). The cells demonstrated marked pleomorphism with hyperchromasia, abundant eosinophilic cytoplasm, and prominent nucleoli. The tumor cells contained both typical and atypical mitoses with an index of 1–4 mitoses per high-power field. Multiple giant cells, scattered acute and chronic inflammation, ectatic capillary vessels, and focal hemorrhage were present. Perineural infiltration, lymphovascular invasion, and necrosis were not identified in the planes of section examined. Both tumors were



FIGURE 1. A 3.5-cm pink, pearly irregularly shaped plaque was noted over the right temple, with light brown crust along the edges and a hyperpigmented focus at the inferior aspect.

transected laterally and at the base. The classic BCC tumor cells were diffusely and strongly positive for Ber-EP4 (Fig. 6), whereas the spindle cells were diffusely and strongly immunoreactive for CD10 (Fig. 7). Based on the histologic and immunohistochemical findings, the case was diagnosed as a collision tumor comprised of AFX

and BCC. Because of the fact that the tumor was broadly transected at the base, the more aggressive undifferentiated pleomorphic sarcoma (uPS) could not be excluded.

Because of the location of the lesion, morphology, and ill-defined margins, the decision was made to proceed with a Mohs excision. The

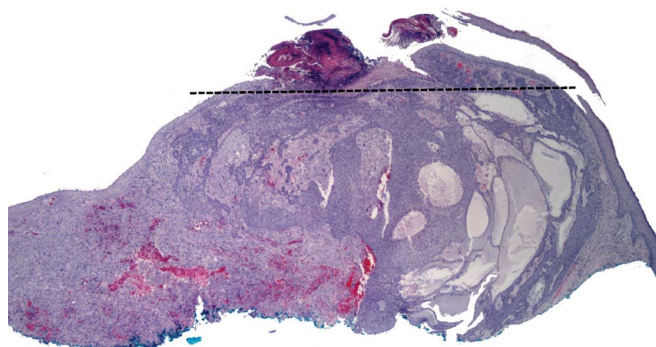


FIGURE 2. A shave biopsy of skin consisting of 2 histologically distinct tumors. There is a cystic basaloid proliferation on the right and a spindle cell proliferation on the left. The area above the dashed line represents what would be viewed on a more superficial shave biopsy ($\times 2$ objective).

The authors declare no conflicts of interest.

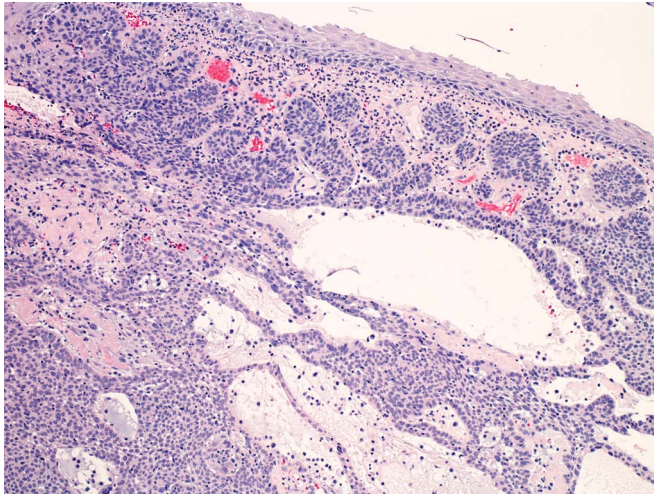


FIGURE 3. There is a basaloid proliferation of cells with nodular and cystic morphology, surrounded by a mucinous stroma ($\times 10$ objective).

residual BCC component of the tumor extended into the subcutis and was completely excised with no complications.

There was no residual AFX component identified in the Mohs sections. Given the patient's complicated dermatologic

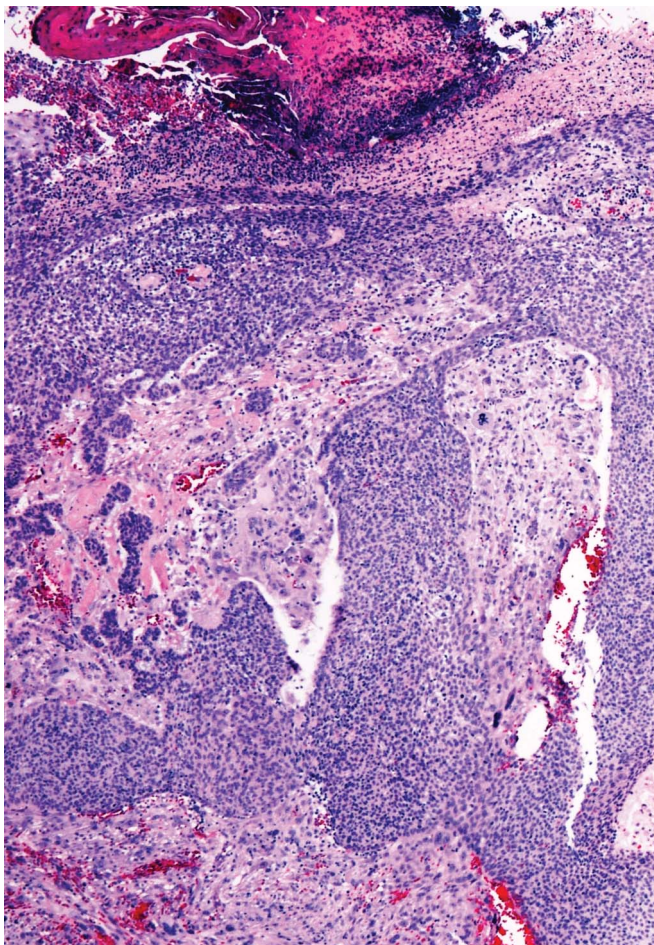


FIGURE 4. Adjacent to and abutting this component is a pleomorphic and mitotically active spindle cell proliferation ($\times 10$ objective).

Copyright © 2015 Wolters Kluwer Health, Inc. All rights reserved.

history, he will require at least annual follow-up.

A collision tumor is defined as 2 or more histologically distinct neoplasms coexisting in the same anatomical location with clearly defined boundaries, and can pose clinical and histologic diagnostic challenges. This is especially true when they involve a combination of 2 malignant tumors, each with its own prognosis, treatment, and ability to metastasize, which cause additional risk to the patient if misdiagnosed. Although the exact etiology of collision tumors is unknown, most authors believe that these combinations occur serendipitously, either because of the high biopsy rate of the 2 tumors or the high incidence of both tumors in sun-exposed skin. However, others suggest that epithelial or stromal changes in 1 tumor can induce the formation of the other,^{1,2} or that both are derived from similar cell lineages.³

AFX is a rare component of collision tumors, with combinations including primarily malignant entities, such as Merkel cell carcinoma,⁴ squamous cell carcinoma in situ,⁵ and invasive melanoma.^{6,7} Although it is generally classified as a low-grade sarcoma, its clinical behavior remains controversial, with outcomes ranging from spontaneous regression⁸ to recurrence and distant metastatic disease,^{9,10} especially after incomplete excision.¹¹ Because of the high rate of local recurrence and ability for metastatic disease, wide local excision with 1-cm margins or Mohs micrographic surgery are recommended.^{12,13} Many believe that AFX is a more superficial and less aggressive variant of uPS, for which treatment requires margins up to 4 cm in some reports.¹⁴

BCCs are common components of cutaneous collision tumors, with the most common combinations, in contrast to AFX, being with benign entities, such as melanocytic nevus, seborrheic keratosis, and neurofibroma.¹⁵ Because of the indolent course and extremely low incidence of metastases (0.0028%–0.55%)¹⁶ in BCC, they are often effectively managed by electrodesiccation and curettage or local excision with 4-mm margins.

We present this case because the combination of BCC and AFX is a rare finding in collision tumors, with only

www.amjdermatopathology.com | 951

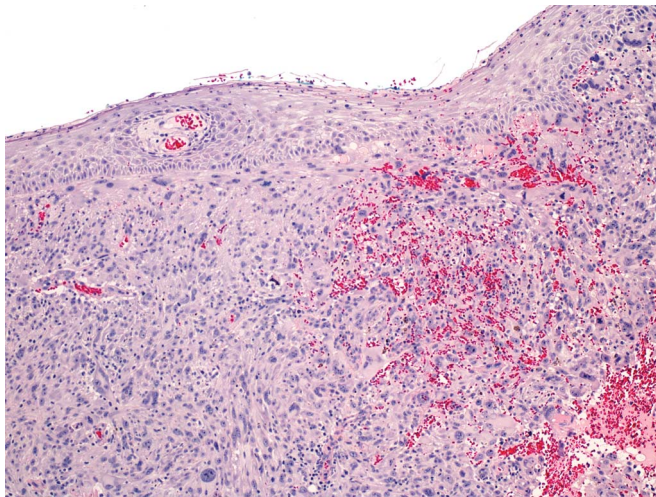


FIGURE 5. There is an infiltrating spindle cell proliferation in a haphazard arrangement that abuts but is not connected to the overlying epidermis. Marked pleomorphism and multiple atypical mitoses are present (×10 objective).

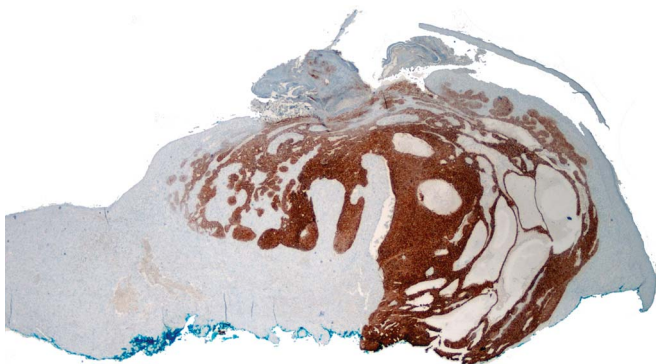


FIGURE 6. Strong and diffuse Ber-EP4 positivity in the basaloid tumor component (×2 objective).

1 other case reported in the literature without discussion of possible therapeutic differences. Although the AFX component in our case represented

a large portion of the biopsy, it is important that dermatopathologists be aware of this potential combination. If a more superficial shave biopsy had

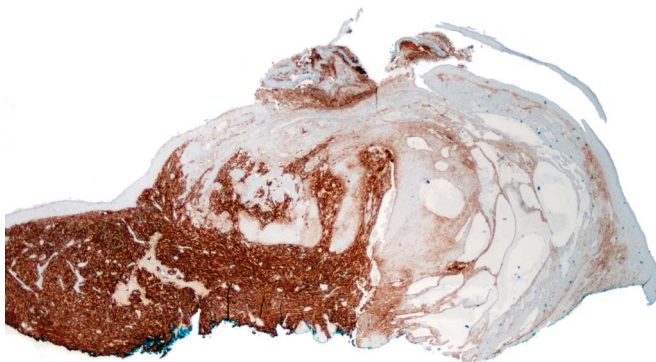


FIGURE 7. Strong and diffuse CD10 positivity in the spindle cell tumor component (×2 objective).

been performed on this patient (demonstrated by area above dashed line in Fig. 2), it could have potentially lead to misdiagnosis of a focus of invasive BCC. The higher incidence of local recurrence and possibility of distant metastases for AFX requires a wider surgical margin and more attentive clinical follow-up. Additionally, a focus of AFX may represent the superficial portion of the more aggressive uPS, which would significantly alter the patient’s treatment and prognosis. We report this collision tumor as an interesting twist on a “routine” case of BCC.

Jodi J. Speiser, MD*
Smitha Aggarwal, MD†
Lindsay Wold, BS†
Rebecca Tung, MD†
Kelli A. Hutchens, MD*

Departments of *Pathology, and †Dermatology, Loyola University Medical Center, Maywood, IL

REFERENCES

1. Brownstein MH, Starink TM. Desmoplastic trichoeplithelioma and intradermal nevus: a combined malformation. *J Am Acad Dermatol.* 1987;17:489–492.
2. Rosenblum GA. Large basal cell carcinoma in a congenital nevus. *J Dermatol Surg Oncol.* 1986;12:166–168.
3. Burkhalter A, White WL. Malignant melanoma in situ colonizing basal cell carcinoma. A simulator of invasive melanoma. *Am J Dermatopathol.* 1997;19:303–307.
4. Youker SR, Billingsley EM. Combined Merkel cell carcinoma and atypical fibroxanthoma. *J Cutan Med Surg.* 2005;9:6–9.
5. Hyatt AM, Mutasim DF, Spicknall KE. Collision of atypical fibroxanthoma and acantholytic squamous cell carcinoma in situ. *Am J Dermatopathol.* 2012;34:563–564.
6. Wilsher MJ. Collision tumor: atypical fibroxanthoma and invasive melanoma. *Pathology.* 2009;41:699–701.
7. McGregor DH, Cherian R, Romanas MM, et al. Amelanotic malignant melanoma: two collision tumors presenting as basal cell carcinoma and atypical fibroxanthoma. *Ann Clin Lab Sci.* 2008;38:157–162.
8. Stefanato CM, Robson A, Calonje JE. The histopathologic spectrum of regression in atypical fibroxanthoma. *J Cutan Pathol.* 2010;37:310–315.
9. New D, Bahrami S, Malone J, et al. Atypical fibroxanthoma with regional lymph node metastasis: report of a case and review of the literature. *Arch Dermatol.* 2010;146:1399–1404.
10. Al Diffalha S, Patel AK, Wojcik EM, et al. Two cases of metastasizing AFX without histologic clues on superficial biopsy [abstract]. *Mod Pathol.* 2013;26(suppl 2). The 102nd

- USCAP Annual Meeting Abstracts; March 2–8, 2013; Baltimore, MD.
- Strutton G, Rubin AI, Weedon D. Tumors and tumor-like proliferations of fibrous and related tissues. In: Strutton G, Rubin AI, Weedon D, eds. *Weedon's Skin Pathology*. China: Elsevier; 2011:Chapter 34, page 836.
 - Giuffrida TJ, Kligora CJ, Goldstein GD. Localized cutaneous metastases from an atypical fibroxanthoma. *Dermatol Surg*. 2004;30(12 pt 2):1561–1564.
 - Ang GC, Roenigk RK, Otley CC, et al. More than 2 decades of treating atypical fibroxanthoma at mayo clinic: what have we learned from 91 patients? *Dermatol Surg*. 2009;35:765–772.
 - Graeber GM. Chest wall resection and reconstruction. *Semin Thorac Cardiovasc Surg*. 1999;11:251–263.
 - Boyd AS, Rapini RP. Cutaneous collision tumors. An analysis of 69 cases and review of the literature. *Am J Dermatopathol*. 1994;16:253–257.
 - Wysong A, Aasi SZ, Tang JY. Update on metastatic basal cell carcinoma: a summary of published cases from 1981 through 2011. *JAMA Dermatol*. 2013;149:615–616.

seasonal allergies, and she denied a history of acne, trauma, or similar lesions in family members. Physical examination showed flesh-colored, 2–4 mm diameter nonfollicular papules scattered diffusely on her shoulders, upper back, and midback (Figs. 1A, B). No surrounding erythema, pustules, plaques, or scales were seen. Eruptive collagenomas, acne scars, and anetoderma were the initial differential diagnoses. A 4-mm punch biopsy was performed, and histopathologic evaluation revealed an essentially normal epidermis. Focally in the mid and reticular dermis, the collagen bundles were miniaturized, appearing both shorter and thinner than adjacent fibers and arranged in a more compact fashion. A Verhoeff-Van Geison stain showed fragmented elastic fibers amidst the miniaturized collagen (Figs. 2A, B). Colloidal iron stain was negative for the presence of

mucin. Screening radiographs of hands and knees were performed and yielded no evidence of osteopoikilosis, sclerosis, or striated epiphyses, all findings would suggest an association with Buschke–Ollendorff syndrome. Complete blood count and chemistry panels were normal, as well as liver enzymes, antinuclear antibody and antiphospholipid antibodies. The patient was prescribed tretinoin 0.04% microgel for daily use over the affected areas. She has had no response to treatment and no progression of disease to date.

Our case is histologically and clinically most consistent with the diagnosis of PE with a unique feature of collagen miniaturization that has never before been reported. The diagnosis of PE can be challenging because of the heterogeneous group of elastic tissue disorders and connective tissue nevi. Additionally, although PE can be

A Pruritic Hypopigmented Cutaneous Eruption: Atypical Presentation of Papular Elastorrhexis

To the Editor:

Papular elastorrhexis (PE) is a rare disorder of elastic tissue that is infrequently reported in the literature. We present a case of a 29-year old Asian woman who presented with a 2-month history of an eruption of asymptomatic flesh-colored papules on her posterior neck and upper back. Her medical history included eczema and

The authors declare no conflicts of interest.

The views expressed in this article are those of the author and do not necessarily reflect the official policy or position of the Department of the Navy, Department of Defense, or the United States Government. I am a military service member. This work was prepared as part of my official duties. Title 17, USC, § 105 provides that “Copyright protection under this title is not available for any work of the United States Government.” Title 17, USC, § 101 defines a US Government work as a work prepared by a military service member or employee of the US Government as part of that person’s official duties.

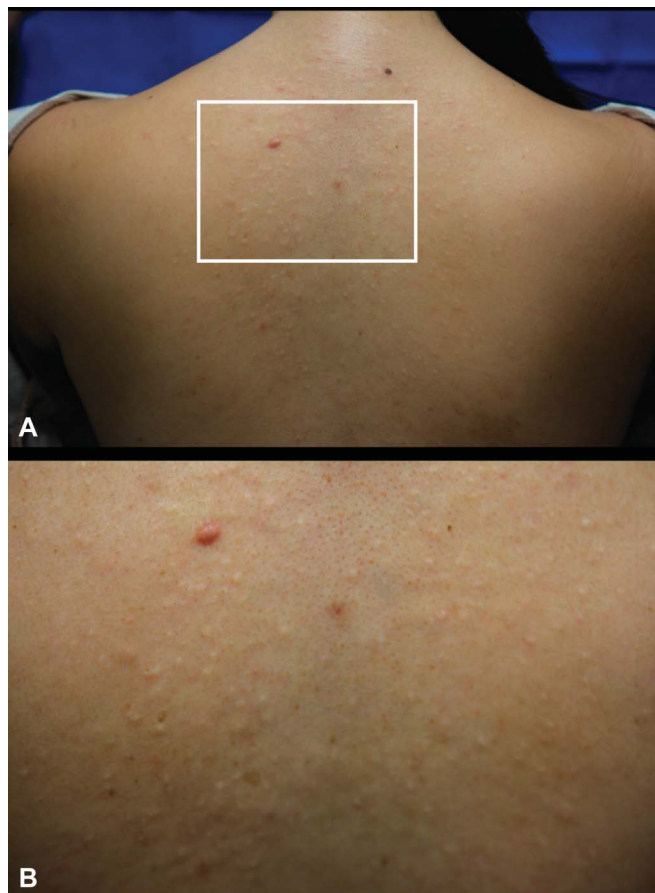


FIGURE 1. A, Photograph of hypopigmented papules distributed across the upper back. B, Inset of a close-up view of papules.