# Concomitant Fibrofolliculoma and Trichodiscoma on the Abdomen

Jessica Riley, DO; Leela Athalye, DO; Donna Tran, DO; Stephanie Fogelson, MD; Paul Shitabata, MD

## PRACTICE **POINTS**

- Fibrofolliculoma and trichodiscoma are flesh-colored adnexal tumors that arise from or around hair follicles.
- It is important to recognize these entities, as they can be related to Birt-Hogg-Dubé syndrome.

Fibrofolliculoma and trichodiscoma are adnexal tumors that arise from or around hair follicles and are two of the many characteristic features of Birt-Hogg-Dubé (BHD) syndrome. Fibrofolliculoma and other hair follicle hamartomas can be differentiated from their clinically indistinct counterparts (eg, trichodiscomas, trichoadenomas) by histologic and staining comparison. We report a rare case of a 54-year-old man who presented with a subcutaneous papule on the abdomen that was histologically proven to have features of both a solitary fibrofolliculoma and trichodiscoma.

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Fibrofolliculomas and trichodiscomas typically present on the head or neck as smooth, flesh-colored, dome-shaped papules. These two entities are considered to constitute two separate time points on a spectrum of histopathologic changes in mantleoma differentiation.[1] Histologically, both are benign hamartomas of the pilosebaceous subunit and collectively are known as mantleomas. We present an unusual case of a concomitant fibrofolliculoma and trichodiscoma on the abdomen.

## Case Report

An asymptomatic 54-year-old man presented for a routine full-body skin examination. A solitary, 2×1-cm, subcutaneous, doughy, mobile nodule was found on the left side of the abdomen with an overlying 2-mm yellow fleshy papule. The patient declined excision of the lesion, and it was recommended that he return for follow-up 3 months later.

The patient did not present for follow-up until 4.5 years later, at which point the lesion had grown to 3.0×2.5 cm in size. An excision was performed, at which time the lesion was noted to be cystic, extruding an oily, yellow-white liquid. Bacterial culture was negative. Histopathologic sections showed a dome-shaped papule with connection to the overlying epidermis. Epithelial extensions from the infundibular epithelium formed a fenestrated pattern surrounding a fibrous and mucinous stroma (Figure, A and B). The differential diagnosis at this time included an epidermal inclusion cyst, fibroma, intradermal nevus, verruca, hemangioma, angiofibroma, and lipoma.[2-4]

The same lesion cut in a different plane of sectioning showed an expansile dermal nodule comprising clusters of sebaceous lobules surrounding a fibrous and mucinous stroma. Within the second lesion, fibrous and stromal components predominated over epithelial components (Figure, C). A diagnosis of fibrofolliculoma showing features of a trichodiscoma arising in the unusual location of the abdomen was made.

## Comment

Solitary fibrofolliculomas and trichodiscomas are flesh-colored, dome-shaped papules that generally present on the face, specifically on the chin, nose, cheeks, ears, and eyebrows without considerable symptoms.[2,4,5] Clinically, fibrofolliculomas are indistinguishable from trichodiscomas but demonstrate different features on biopsy.[1,5]

Fibrofolliculomas and trichodiscomas are well known for their association with Birt-Hogg-Dubé (BHD) syndrome when they present concomitantly and typically arise earlier in the third decade of life than solitary

Dr. Riley is from the Department of Dermatology, St. Joseph Mercy Health System, Ann Arbor, Michigan. Drs. Athalye and Tran are from Western University of Health Sciences, Pomona, California, and the Department of Dermatology, College Medical Center, Long Beach, California. Dr. Fogelson is from the Dermatology Center at Ladera, Ladera Ranch, California. Dr. Shitabata is from the Department of Dermatology, Harbor-UCLA Medical Center, Torrance, California.

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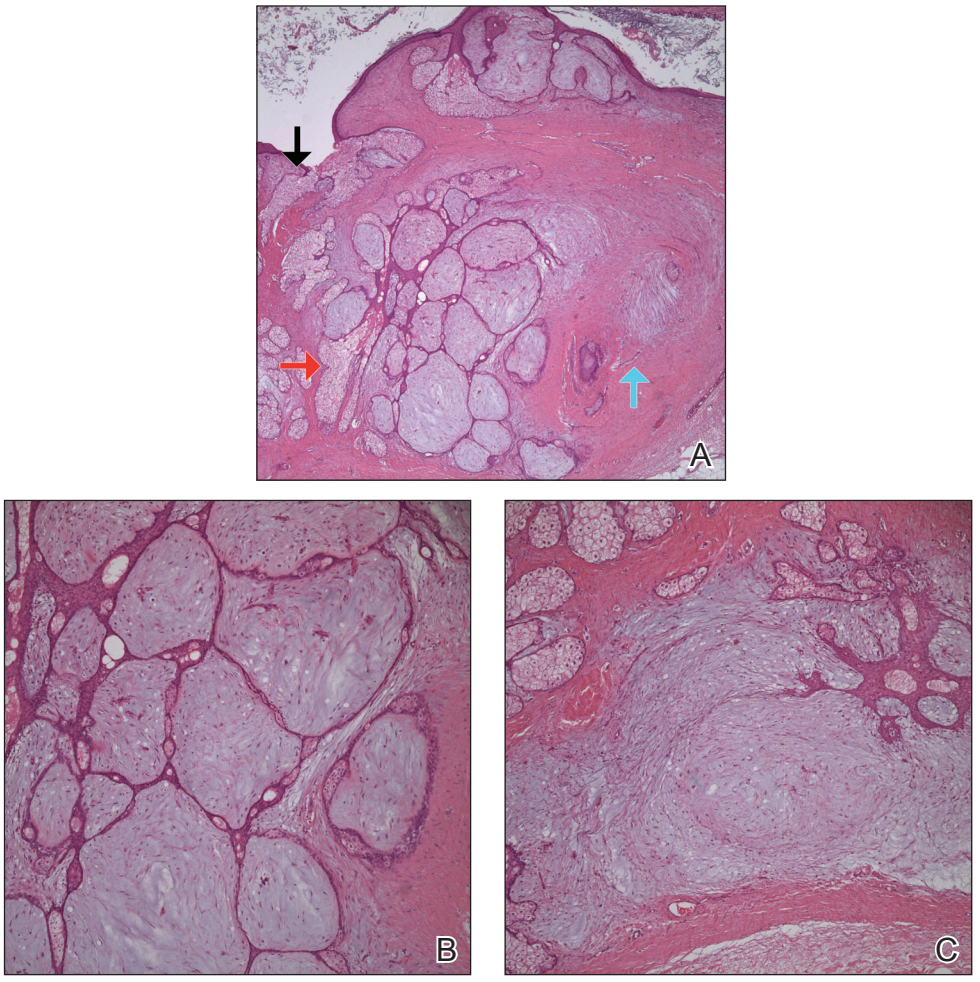
Correspondence: Jessica Riley, DO, 1300 Avenida Vista Hermosa, Ste 150, San Clemente, CA 92673 (drjessicariley@gmail.com).




An epidermal inclusion cyst (black arrow) arising in association with a fibrofolliculoma (red arrow) and exhibiting features of a trichodiscoma (blue arrow)(H&E, original magnification ×20). The fibrofolliculoma component is characterized by thin epithelial strands composed of thin cords of bland epithelial cells surrounding loose stroma with bland spindle cells and mucin (B)(H&E, original magnification ×40). The trichodiscoma component demonstrated an expansile dermal nodule comprised of clusters of sebaceous lobules surrounding a fibrous and mucinous stroma (C)(H&E, original magnification ×20).

fibrofolliculomas; however, there have been reports of solitary fibrofolliculomas in patients aged 1 to 36 years.[4,6] The triad of BHD syndrome consists of multiple fibrofolliculomas, trichodiscomas, and acrochordons, and it is acquired in an autosomal-dominant manner, unlike solitary fibrofolliculomas, which typically are not inherited. Birt-Hogg-Dubé syndrome is caused by a mutation in the *FLCN* gene that codes for the tumor-suppressor protein folliculin, which when mutated can cause unregulated proliferation of cells.[7] Solitary fibrofolliculomas and the multiple fibrofolliculomas seen in BHD syndrome are histologically similar.

Fibrofolliculoma can be clinically indistinguishable from fibroepithelioma of Pinkus, perifollicular fibroma, trichilemmoma, trichodiscoma, trichoepithelioma, and trichofolliculoma. All typically present clinically as flesh-colored papules,[1] although histologic distinction can be made (Table).[5,8-13]

Fibrofolliculoma is a benign hamartoma that arises from the pilosebaceous follicle and consists of an expansion of the fibrous root sheath, which typically surrounds the hair follicle along with proliferating bands or ribbons of perifollicular connective tissue. As such, the hair follicle may be dilated and filled with keratin in the expanded infundibulum.[8] Follicles also may be surrounded by a myxoid stroma.[2] In contrast, trichodiscoma is characterized by connective tissue with mature sebaceous lobules in the periphery. It has a myxoid stroma, as opposed to the more fibrous stroma seen in fibrofolliculomas.

Reports have examined the staining patterns of fibrofolliculomas, which show characteristics similar to those of other hair follicle hamartomas, including trichodiscomas.[10] The connective tissue and epithelial components that constitute a fibrofolliculoma show different staining patterns. The connective tissue component stains positive for CD34 spindle cells, factor XIIIa, and nestin (a marker

**Differentiating Histopathologic Features of Adnexal Derivatives**[5,8-13]

| Adnexal Entity | Distinguishing Histopathologic Features |
|---|---|
| Fibrofolliculoma | Dilated, central follicular infundibulum; epithelial strands of basaloid cells emanating from the infundibulum of the hair follicle |
| Fibroepithelioma of Pinkus | Strands of palisading basaloid cells contiguous with the epidermis; embedded in stroma |
| Perifollicular fibroma | Circular arrangement of fibrous tissue surrounding a hair follicle; absence of mature sebaceous lobules |
| Trichilemmoma | Lobules of keratinocytes in the epidermis; peripheral palisading; eosinophilic basement membrane |
| Trichodiscoma | Proliferation of connective tissue and fibrous stroma; located near a hair follicle |
| Trichoepithelioma | Islands of basaloid cells aggregating in a concentric manner; mesenchymal bodies in the papillary dermis |
| Trichofolliculoma | Lobules of basaloid cells emanating from a dilated infundibulum; shows all stages of follicle differentiation including secondary hair follicles and rudimentary hair bulb |

of angiogenesis). CD117 (c-kit) expression in the stroma, a marker of fibrocytes, is a feature of both fibrofolliculoma and perifollicular fibromas. The epithelial component, consisting of the hair follicle itself, stains positive for CK15. CK15 expression has been reported in undifferentiated sebocytes of the mantle and in the hair follicle.[10] Immunohistochemical staining supports the notion that fibrofolliculomas contain connective tissue and epithelial components and helps to compare and contrast them to those of other hair follicle hamartomas.

Ackerman et al[1] considered both fibrofolliculomas and trichodiscomas to be hamartomas of the epithelial hair follicle. The exact etiology of each of these hamartomas is unknown, but the undifferentiated epithelial strands protruding from the hair follicle in a fibrofolliculoma lie in close proximity to sebaceous glands. Furthermore, the authors postulated that fibrofolliculomas and trichodiscomas constitute a spectrum that encompasses the differentiation process of a mantleoma, with fibrofolliculoma representing the beginning of mantleoma differentiation and trichodiscoma representing the end. This end stage of follicular differentiation is one in which there is a predominant stroma and the previously undifferentiated epithelium has formed into sebaceous ducts and lobules in the stroma.[1]

Most cases of fibrofolliculoma and/or trichodiscoma arise in areas of dense sebaceous follicle concentration (eg, face), further supporting the hypothesis that sebaceous gland proliferation contributes to fibrofolliculoma.[14] The case described here, with the fibrofolliculoma arising on the abdomen in conjunction with a trichodiscoma, is therefore worth noting because its location differs from what has been observed in previously reported cases.[4]

There are both surgical and medical options for treatment of fibrofolliculoma. Although surgical excision is an option for a single lesion, patients with multiple fibrofolliculomas or BHD may prefer removal with the combined $CO_2$ laser and erbium-doped YAG laser.[15]

## Conclusion

We present a rare case of concomitant fibrofolliculoma and trichodiscoma arising on the unusual location of the abdomen. This report highlights the histopathologic features of multiple adnexal tumors and emphasizes the importance of biopsy for differentiating fibrofolliculoma and trichodiscoma.

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