Solitary Nasal Fibrofolliculoma

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Citation

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Abstract

Fibrofolliculoma is a very rare benign tumor of the skin that is derived from the perifollicular sheath. Histologically, it shows a well-formed central hair follicle with a dilated infundibulum containing laminated keratin with anastomosing epithelial strands that radiate from the central hair follicle into the perifollicular fibrotic stroma. The patients with multiple fibrofolliculomas have an association with Birt-Hogg-Dube syndrome. We report a case of a solitary nasal fibrofolliculoma occurring in a 60-year-old male and briefly review the literature.

INTRODUCTION

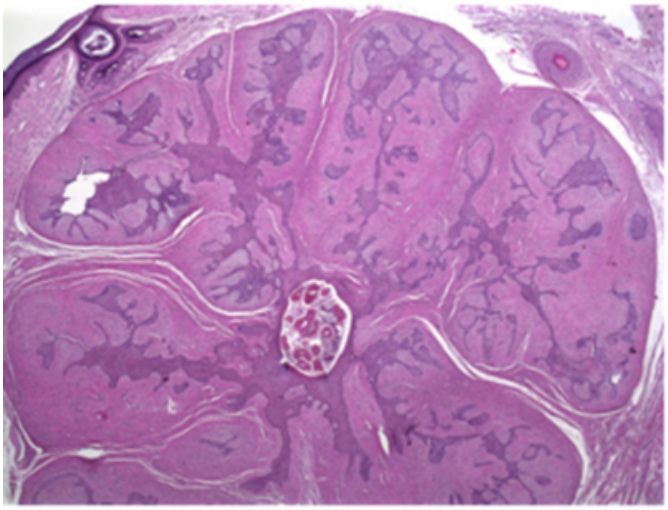
Fibrofolliculoma is a very rare benign follicular tumor of the skin that may be a variant of perifollicular fibroma and trichodiscoma [1,2]. The patients with multiple fibrofolliculomas are associated with Birt-Hogg-Dube syndrome that may include renal carcinoma, spontaneous pneumothorax, and colonic adenocarcinoma [3]. We are reporting a case solitary nasal fibrofolliculoma and briefly review the literature.

CASE REPORT

A 60-year-old male presented with a nasal nodule present for an unknown period of time. An excisional biopsy revealed a 1.2-cm well-circumscribed dermal mass with a white-tan cut surface. Microscopically, the epidermis was normal. The dermis revealed a well-circumscribed tumor nodule with a central hair follicle (Figure 1).

Figure 1

Figure 1: Fibrofolliculoma, low magnification.



The hair follicle was well formed with a dilated infundibulum containing laminated keratin. The basaloid epithelial cells formed interconnecting and anastomosing strands that radiated from the central hair follicle. The perfollicular stroma consisted of dense fibrotic tissue with focal areas of loose myxoid changes (Figure 2). The tumor cells were monotonously small with round, oval dark nuclei and inconspicuous nucleoli (Figure 3).

Additional history and clinical studies did not reveal any other skin tumors or any internal neoplasms.

Figure 2

Figure 2: Fibrofolliculoma, higher magnification.

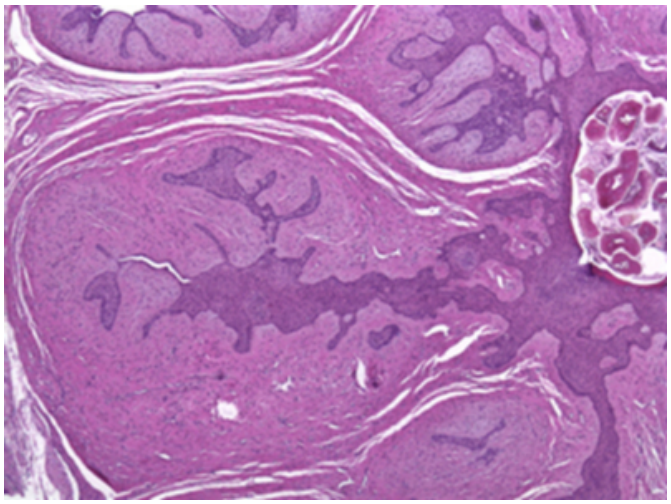
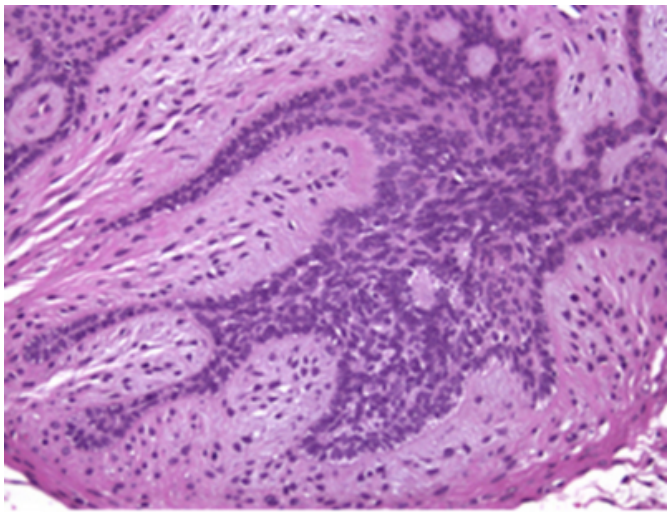


Figure 3

Figure 3: Fibrofolliculoma, higher magnification.



DISCUSSION

Fibrofolliculoma is a very rare benign tumor of the skin that is derived from the perifollicular sheath. The tumor commonly presents as asymptomatic, multiple, small, white or flesh-colored, smooth, dome-shaped papules predominately over the scalp, face, oral cavity, neck, and upper trunk. Microscopically, the tumor shows a well-circumscribed dermal tumor with a central well-formed dilated hair follicle. The neoplastic epithelial cells form thin anastamosing cords that radiate from the central hair follicle. The perifollicular stroma is accentuated and fibrotic.

Patients with multiple fibrofolliculomas have an association with Birt-Hogg-Dube syndrome (BHDS) that presents with cutaneous fibrofolliculomas, trichodiscomas, and acrochordons [3]. Trichodiscoma is a small hamartomatous tumor of the hair disk with a proliferation of the fibrovascular component of the hair disk and small melanin granule containing cells and occasional myelinated nerves. Perifollicular fibroma characteristically has a central hair follicle with hair shaft that is surrounded by circumferentially arranged fibrous sheath. Fibrofolliculoma, trichodiscoma, and perifollicular fibroma are closely related lesions. They may be the same entity and the different histologic appearances may be due to the plane of sectioning.

BHDS is also associated with renal tumors, particularly chromophobe renal carcinoma and renal oncocytoma, pulmonary cysts, spontaneous pneumothorax, colonic polyps, and colonic carcinoma [5]. Other associated features include large connective-tissue nevus, parathyroid adenomas, flecked chorioretinopathy, bullous emphysema, lipomas, angiolipomas, parotid oncocytomas, multiple oral mucosal papules, neural tissue tumors, and multiple facial angiofibromas. BHDS has an autosomal dominant inheritance with a mutation on band 17p11.2 that involves a novel BHDS protein called folliculin [4, 6].

Additional clinical history, careful physical examination, and screening tests may berequired for a patient with a histologic diagnosis of fibrofolliculoma to exclude possible BHDS and associated neoplasms.

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