Nail psoriasis is seen in 50% of patients with psoriasis and in about 80% of patients with psoriatic arthritis. The management of nail psoriasis can be very challenging. The choice of treatment depends on the clinical presentation and specific patient factors. Systemic treatment is indicated when nail psoriasis is severe or extensive skin lesions and psoriatic arthritis coexist or when topical treatment fails.

In our patient, ustekinumab proved rapidly efficacious in the treatment of nail disease, improving his nail abnormality-induced psychological stress. Our findings are in agreement with existing limited data regarding the effectiveness of ustekinumab in nail psoriasis.

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**Multiple Desmoplastic Melanomas in Birt-Hogg-Dubé Syndrome and a Proposed Signaling Link Between Folliculin, the mTOR Pathway, and Melanoma Susceptibility**

Birt-Hogg-Dubé syndrome (BHDS) is a rare autosomal dominant disorder characterized by the development of cutaneous and systemic tumors. The BHD gene (OMIM 135150) codes for the protein folliculin, which is expressed in multiple tissues including kidney, lung, and skin. Folliculin is thought to play a role in tumor suppression exerting an inhibitory effect on the growth-promoting mammalian target of rapamycin (mTOR) pathway. Mutations observed within the BHD gene in BHDS lead to the expression of inactive folliculin, ultimately resulting in mTOR pathway activation. Unregulated mTOR activation promotes cell growth and proliferation, as observed in melanoma pathogenesis.

**Report of a Case.** A 58-year-old man was seen for multiple, histologically proven, facial fibrofolliculomas (Figure 1A and B) raising the possibility of BHDS. He had a history of spontaneous pneumothoraces in his third
A decade of life and a left submandibular salivary oncocytoma in 2002. We confirmed BHDS via detection of a previously reported, heterozygous, truncating, insertional mutation within the BHD gene (c.1285_1286insC).\textsuperscript{2} Renal tract ultrasonography and chest, abdomen, and pelvis computed tomographic findings were unremarkable.

Twenty-one years earlier, he developed a desmoplastic melanoma on his right cheek (Breslow thickness, 0.5 mm; Clark level, 3) requiring a regional lymphadenectomy for cervical lymph node metastases 3 years later. In 2006, he developed a lentigo maligna on his right cheek at the site of his primary desmoplastic melanoma. In June 2009, he developed a second desmoplastic melanoma (Breslow thickness, 2.4 mm; Clark level, 4) on his left shoulder (Figure 1C).

Comment. This patient developed 2 desmoplastic melanomas, which are an uncommon, locally aggressive variant representing 1% to 3% of melanomas. Melanoma has been previously reported in 6 patients with BHDS,\textsuperscript{1,2,5} although none had multiple or recurrent lesions. The BHD gene mutations in 4 of these 6 occurred within a mutational “hot spot” in a highly conserved 700-kb region of the BHD gene on chromosome 17p11.2.\textsuperscript{1,2}

Though the exact function of the highly conserved BHD gene product folliculin remains unknown, experimental evidence suggests that it has a role in tumor suppression.\textsuperscript{2} Figure 2 illustrates the complex series of interactions between folliculin and the growth-promoting mTOR pathway.\textsuperscript{3} Inactive folliculin, as expressed in BHDS, being unable to bind to active adenosine monophosphate-activated protein kinase (Figure 2), might lead to mTOR activation and uncontrolled cell growth.\textsuperscript{1,2}

Dysregulation of the mTOR pathway is associated with the putative pathogenic pathways of benign and malignant tumors and, more recently, has been considered as a candidate target for melanoma therapy.\textsuperscript{3,4} The mTOR inhibitor rapamycin decreases cell growth and proliferation in vitro and tumor proliferation in vivo.\textsuperscript{5,4} It has yielded encouraging in vitro and in vivo data when used in combination with other cytotoxic agents to treat metastatic melanoma.\textsuperscript{6}

Establishing incidence vs coincidence for malignant associations with rare conditions like BHDS is difficult. While the question of whether mutated folliculin in BHDS predisposes patients to melanoma requires further investigation, it raises the possibility of the potential use of mTOR inhibitors in BHDS as a treatment option for the range of associated malignant neoplasms.\textsuperscript{3,6}

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Localized Porokeratosis Secondary to Ionizing Radiotherapy for Prostate Carcinoma

Segmental porokeratosis developing after electron beam radiotherapy for follicular lymphoma has been reported once, to our knowledge, in the literature.1 However, the patient in that case could not recall which areas of skin were irradiated, so the association was open to question. We report a second case, which we think demonstrates the association even more convincingly.

Report of a Case. Our patient was diagnosed as having prostate adenocarcinoma and treated with bicalutamide, leuprorelin, and radiotherapy (total radiation dose, 64 Gy in 32 fractions over 6 weeks) administered as 1 anterior and 2 lateral fields. (To convert Grays to rads, multiply by 100.) The anterior field size was 9.0 × 8.6 cm. Treatment was completed 7 months after diagnosis.

He was seen by us 6 years later, at age 76 years, and had multiple erythematous lesions with raised scaly margins localized to the natal cleft and medial aspects of both buttocks (Figure 1). Skin biopsy specimens (Figure 2) showed several angulated columns of parakeratin with underlying hypogranulosis and dyskeratosis (cornoid lamellae), consistent with a diagnosis of porokeratosis. Liquid nitrogen cryotherapy successfully cleared the lesions.

Radiotherapy mapping images confirmed that the area of porokeratosis coincided exactly with the exit dose of the anterior radiation field (Figure 3). The dose received by the skin over the natal cleft was calculated at 28% of the applied dose, ie, 18 Gy, delivered in 32 fractions of 0.56 Gy.