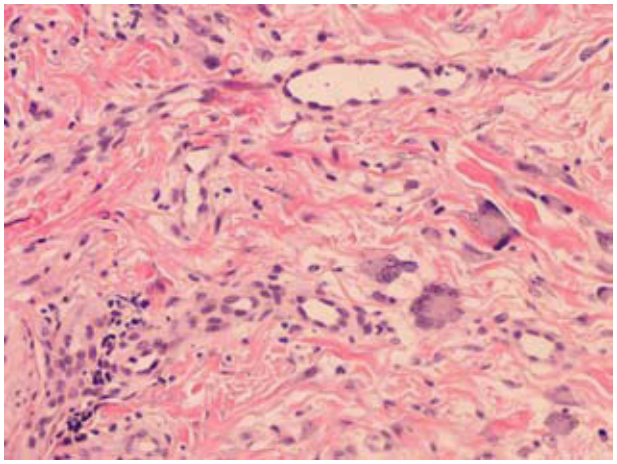

Figure 2 Image of the dermis showing vascular proliferation and an infiltrate formed of mixed cells and multinucleated giant cells with angulated cytoplasm (hematoxylin-eosin, original magnification ×100).

The generalized variant of MCA is extremely rare, with only 3 cases recorded in the literature, to which we add a new case that is not associated with any other disease.

Conflict of Interest

The authors declare that they have no conflict of interest.

Acknowledgments

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Simultaneous Presentation of Localized Basaloid Follicular Hamartoma and Epithelioid Blue Nevus in a 44-Year-Old Patient

Hamartoma folicular basalioide localizado y nevus azul epitelioide de presentación simultánea en una paciente de 44 años

To the Editor:

Basaloid follicular hamartoma is an uncommon benign neoplasm whose histological appearance is very similar to that of infundibulocystic basal cell carcinoma and trichoepithelioma. It may be familial or sporadic, generalized or localized, and is occasionally associated with autoimmune disease and cystic fibrosis. Some patients have a mutation in the *PTCH* gene that is similar to that observed in Gorlin syndrome and in up to 50% of isolated basal cell carcinomas.

Epithelioid blue nevus is a benign neoplasm whose histological diagnosis is complex due to its similarity with melanocytoma. It is closely associated with Carney complex.

We present the case of a 44-year-old woman with no relevant past history who consulted for a lesion on her back. The woman was unable to say how long she

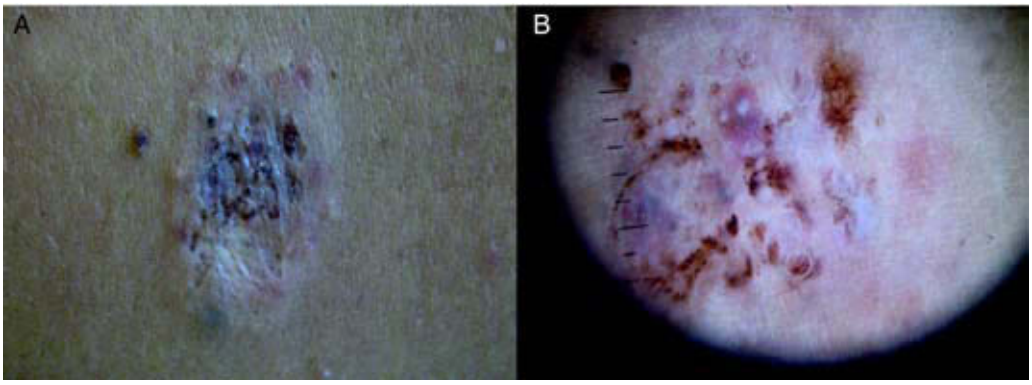

Figure 1 A, Fibrotic plaque on the left scapula. B, Dermoscopic pattern: globular and cobblestone pattern with deep whitish, pinkish, and grayish areas, keratotic plugs, and milia.

had had the lesion. The results of a biopsy taken at another center indicated a histological diagnosis of nevus. Examination revealed a whitish fibrous plaque with a scar-like appearance measuring 2.5 cm in diameter on the left scapula. Dermoscopy revealed a globular pattern with a cobblestone appearance and deep whitish, pinkish, and grayish areas, as well as keratotic plugs and milia (Figure 1). The patient also presented a pedunculated bluish lesion on the left side of the neck. The lesion was asymptomatic and, again, the patient did not know how long she had had it. Both lesions were removed.

Histopathology of the back lesion revealed an epithelial and basaloid proliferation in the superficial and middle dermis that formed solid nests and cords with follicular differentiation and keratotic cysts (Figure 2). The neck lesion was a dermal proliferation of intensely pigmented ovoid and fusiform melanocytic cells arranged in nests and cords and located close to the cutaneous adnexal structures within a scleral stroma (Figure 3). These findings led us to diagnose basaloid follicular hamartoma and epithelioid blue nevus, respectively.

Basaloid follicular hamartoma is an uncommon benign neoplasm of the hair follicle. It was first described by Brown in 1969.[1] There are 5 forms of presentation:

1. Generalized acquired. This type of lesion is sometimes associated with autoimmune diseases such as systemic lupus erythematosus and myasthenia gravis. The lesions are found mainly on the face and periorificial areas.
2. Generalized congenital hereditary. This lesion may be associated with other ectodermal defects such as hypotrichosis and punctate keratotic pits on the palms and soles and with cystic fibrosis.
3. Generalized familial, no associations. This type appears on the face and genital region.
4. Linear unilateral. This type usually follows the Blaschko lines. It may be present at birth or develop during childhood.
5. Localized. Localized basaloid follicular hamartoma was first described in 1992 and is usually found on the scalp.

Histologically, all the clinical forms of basaloid follicular hamartoma include similar findings consisting of affected follicles replaced by epithelial cords 2 or 3 cells thick that emerge from the follicular axis and are sometimes arranged in the form of an inverted candelabrum and small infundibular cysts. The latter may account for the dermoscopic image of keratotic plugs and milia. The stroma surrounding the lesion is poor and fibrotic.

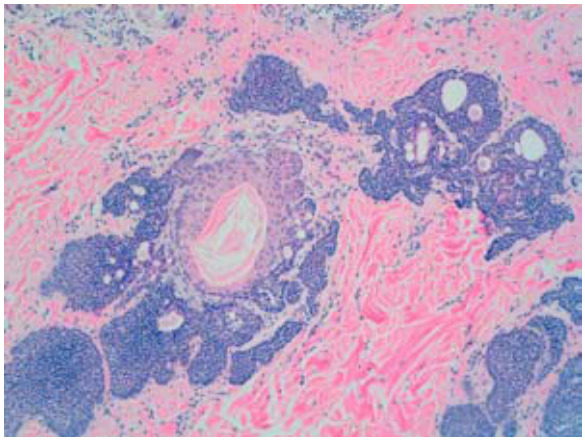

Figure 2 Lesion consisting of nests of basaloid cells and keratotic cysts arranged in the dermis, closely related to the pilosebaceous follicles (hematoxylin-eosin, original magnification ×100).

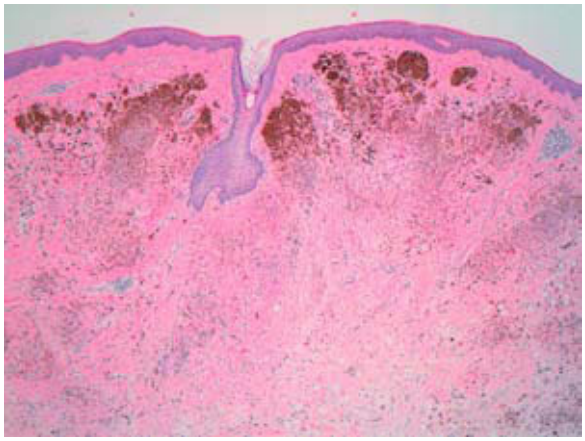

Figure 3 Dermal melanocytic proliferation of epithelioid and fusiform cells, some of which are intensely pigmented, within a collagenous stroma (hematoxylin-eosin, original magnification ×40).

The main differential diagnosis is made with infundibulocystic basal cell carcinoma and trichoepithelioma, both of which are also tumors of the hair follicle. Basaloid follicular hamartoma tends to be more folliculocentric, with less involvement of the interfollicular dermis than infundibulocystic basal cell carcinoma.[2] In cases of incipient infundibulocystic basal cell carcinoma these differences are not as clear, and some authors postulate a possible overlap between the two entities.[3] *PTCH* is a tumor suppressor gene that codes for a transmembrane receptor protein. Mutations in *PTCH* are implicated in the pathogenesis of basaloid epithelial neoplasms, such as trichoepithelioma and basal cell carcinoma. They are present in up to 50% of sporadic basal cell carcinomas and in Gorlin syndrome. As with those tumors, basaloid follicular hamartoma presents mutations in *PTCH*; however, consistent with the benign behavior of these neoplasms, the mutations are different and less common than in the diseases described above.[4] Therefore, a possible relationship between these conditions has been postulated. Another feature of the tumor is that cell proliferation markers such as Ki-67 or proliferating cell nuclear antigen stain basaloid follicular hamartoma less intensely than they do infundibulocystic basal cell carcinoma.

Carney and Ferreiro[5] first described epithelioid blue nevus in 1996 as a component of Carney complex (which includes cardiac and cutaneous myxoma, lentiginosis, endocrine hyperactivity, testicular tumors, and schwannoma), although it can also appear in isolation.[5] Histopathology reveals intensely pigmented epithelioid and fusiform melanocytes with prominent nucleoli. They are sometimes immature at deeper levels and can reach the deep dermis. The differential diagnosis should be made with Spitz nevus and melanocytoma.[6,7] Histological differential diagnosis with the latter is sometimes difficult, although ulceration occurs more frequently in melanocytoma.

Our search of the literature (PubMed, March 2010, key words *epithelioid blue nevus* and *basaloid follicular hamartoma*) revealed no cases of simultaneous basaloid follicular hamartoma and isolated epithelioid blue nevus not associated with Carney complex. For the moment, we do not know whether this association is fortuitous or whether it has wider implications.

To conclude, although basaloid follicular hamartoma shares characteristics with infundibulocystic basal cell carcinoma, its prognosis and treatment are different; therefore, a firm histological diagnosis is important if aggressive treatment of this benign tumor is to be avoided. Epithelioid blue nevus should suggest Carney complex. Histological differentiation between epithelioid blue nevus and melanocytoma is essential, given the malignant potential of the latter.

Finally, the dermoscopic description of basaloid follicular hamartoma is of special interest, since our literature search revealed no previous references.

Conflict of Interest

The authors declare that they have no conflict of interest.

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