Dermatology Practical & Conceptual

Multiple Pigmented Trichoepitheliomas: A Case Report

Stefano Caccavale[1], Carmine Fiorentino[1], Maria Pia Boccellino[1], Francesca Pagliuca[2], Ruzica Jurakic Toncic[3], Giuseppe Argenziano[1]

1 Dermatology Unit, Department of Mental and Physical Health and Preventive Medicine, University of Campania Luigi Vanvitelli, Naples, Italy
2 Division of Pathology, Department of Mental and Physical Health and Preventive Medicine, University of Campania Luigi Vanvitelli, Naples, Italy
3 Department of Dermatology and Venereology, University of Zagreb, Zagreb, Croatia

Citation: Caccavale S, Fiorentino C, Boccellino MP, Pagliuca F, Jurakic Toncic R, Argenziano G. Multiple Pigmented Trichoepitheliomas: A Case Report. *Dermatol Pract Concept*. 2023;13(2):e2023128. DOI: https://doi.org/10.5826/dpc.1302a128

Accepted: November 2, 2022; **Published:** April 2023

Funding: None.

Competing Interests: None.

Authorship: All authors have contributed significantly to this publication.

Corresponding Author: Stefano Caccavale, MD, Dermatology Unit, Department of Mental and Physical Health and Preventive Medicine, University of Campania Luigi Vanvitelli, Via Sergio Pansini, 5, 80131 Naples, Italy; phone: +39 3336365526; fax: +39 0815468759; E-mail: stefano85med@libero.it

Dear Editor,

Trichoepitheliomas are uncommon benign tumors with infundibulocystic differentiation. They most often occur in women, presenting themselves as translucent round papules or nodules mainly localized at the face or scalp [1]. They may appear as solitary or multiple lesions in non-familial or familial forms (including multiple familial trichoepitheliomas, Brooke-Spiegler syndrome and familial cylindromatosis) [1,2]. Familial forms are often due to mutations in CYLD gene, on chromosome 9, with autosomal dominant transmission. This gene has incomplete penetration in males, so the prevalence of familial forms is higher in women. In these cases, the onset of trichoepitheliomas is earlier compared to sporadic forms.

To the best of our knowledge, we report the first case of multiple pigmented trichoepitheliomas discussed in literature so far.

A 70-year-old-man, Fitzpatrick photo-type 4, presented to our attention for Zoon balanitis. During the clinical examination of the total body surface we found multiple nodular pigmented lesions of the face, trunk and lower limbs.

We identified 5 pigmented, non-ulcerated, smooth, brown/blue papules and nodules of 0.5-0.7 cm of diameter. Two lesions were localized on the head, respectively at the superciliary arch and the occiput. The other three lesions were localized at the posterior thigh, the superior back and the chest.

On dermoscopy (Figure 1, A-F), the lesions were highly pigmented and showed basalioma-like features, such as brown-blue globules and nests, blue-black blotches and leaf-like areas together with less specific patterns (shiny-white lines, milia-like cysts and gray-blue veil). Four of them presented bright vessels. The lesion localized at the posterior thigh presented a blue lacunar-like pattern.

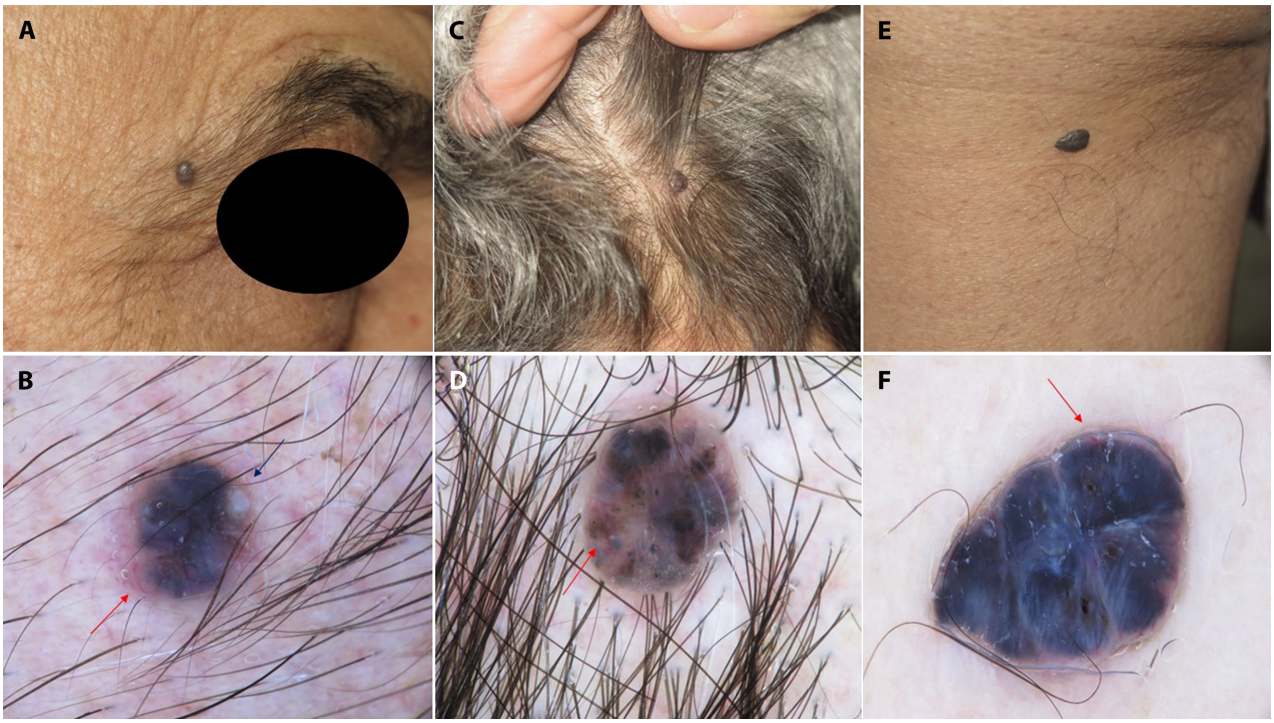

Figure 1. (A-F) Clinical presentation on admission (A,C,E). Dermoscopic images (B,D,F) revealed: bright vessels (red arrows in B,D,F), milia-like cyst (blue arrow in B), leaf-like areas (D), brown-blue globules and nests (D), lacunar-like pattern (F), shiny white lines (F), blue-white veil (B,F), brown-blue blotches (B,F).

Since the dermoscopic clues were suggestive for pigmented basal cell carcinomas, we decided to excise all lesions. The histopathology of the biopsy samples showed islands and nests of follicular germinative cells, without stromal retraction spaces, immersed in a conspicuous fibrocytic stroma, with high cellularity. The well-circumscribed tumors were focal connected with the overlying epidermis. In addition, multiple horn cysts were detected in some lesions. All these features suggested overall the diagnosis of trichoepitheliomas and not of basal cell carcinomas.

Due to the presence of multiple pigmented trichoepitheliomas, a DNA analysis to exclude a possible genetic etiology was proposed to the patient, but he refused.

In conclusion, this was the first case of multiple pigmented trichoepitheliomas described so far in literature to our knowledge. These lesions are clinically and dermoscopically very difficult to distinguish from pigmented basal cell carcinomas, similarly to non-pigmented ones. At date, the histopathological examination still represents the gold standard to the final diagnosis. More studies will be needed to understand if there may be a correlation between the photo-type and predisposition to develop pigmented rather than non-pigmented trichoepitheliomas.

References

1. Karimzadeh I, Namazi MR, Karimzadeh A. Trichoepithelioma: A Comprehensive Review. *Acta Dermatovenerol Croat.* 2018; 26(2):162-165. PMID: 29989874.
2. Supekar BB, Rambhia KD, Tomar SS, Singh RP, Multiple Familial Trichoepithelioma with Varied Malignancies. *Indian J Dermatol.* 2021;66(2):226. DOI: 10.4103/ijd.IJD_647_18. PMID: 34188306. PMCID: PMC8208248.