Unexpected case of non-syndromic neighbouring basal cell carcinomas

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DESCRIPTION

A 66-year-old Caucasian woman (Fitzpatrick skin type III) presented in our department for the evaluation of a lesion in the lumbosacral area, that had first appeared approximately 3 years prior to the referral and was gradually progressing in size. The lesion was a pruritic erythematous and slightly hyperkeratotic papule that, according to the patient, was constantly irritated through contact with clothes, and was occasionally bleeding. During the whole-skin physical examination two other suspicious lesions were documented in a linear distribution in the immediate vicinity of the above-mentioned papule (figure 1). The first presented as an erythematous macula and the second presented as a slightly elevated plaque with an irregular multifocal brown pigmentation in the periphery. The histological examination of the lesions revealed three basal cell carcinomas (BCCs). The tumours were surgically removed via an elliptical excision and their histological examination revealed three basal cell carcinomas (BCCs). The tumors were surgically removed via an elliptical excision and the patient was advised to present for regular skin examination follow-ups, given the recurrence rates of BCCs. The patient was otherwise healthy, without signs of Gorlin-Goltz syndrome or other hereditary disorders, history of malignancy, exposure to carcinogenic substances such as arsenic, topical radiation, solarium treatments or excessive sunbathing. She further denied cases of skin cancer in the family history.

BCC is the most common skin neoplasm, with multiple causes such as ultraviolet (UV) radiation exposure, genetic predisposition with Fitzpatrick skin types I and II, or hereditary conditions, such as Xeroderma pigmentosum and Gorlin-Goltz syndrome. For the cases where numerous BCCs occur without the evident influence of an extrinsic risk factor, the term multiple non-syndromic BCCs is used. The reporting of such patients in the literature is very limited, and the BCCs in the described cases tend to have similar morphological characteristics.

Dysregulated Hedgehog signalling is a hallmark of BCC pathogenesis. The cellular origin of these tumours is believed to arise from the basal cells of the interfollicular epidermis and infundibulum of hair follicles. Although the exact pathogenesis of non-syndromic BCCs is not fully elucidated, an association with the biological events that lead to cutaneous field cancerisation can be speculated. The pathogenetic mechanism of field cancer is based on the following model: acquired genetic alterations in skin stem cells (mostly UV-induced) give origin to proliferating clones that expand in the epidermis and form patches of mutated cells. These clones and subclones acquire additional mutations that can result in carcinomatous transformation over time.

The synchronous appearance of multiple neighbouring BCCs with distinct clinical and dermatoscopic characteristics on areas that are not sun-exposed and in otherwise healthy patients, without history of malignancy or other relevant comorbidities, such as immunosuppression or other pathogenic background, and without relevant family history, is rather uncommon. The peculiar coincidence in our case is the immediate proximity of the three BCCs, their unusual distribution in a...
linear pattern and the fact that they displayed different macroscopic and dermatoscopic features (Figure 2).

The objective of this article is to raise awareness over the existence of non-syndromic BCCs and to point out that physicians should not hesitate to perform diagnostic biopsies in uncertain cases. Clinical suspicion is a factor that should not be underestimated, even in cases of low probability of cutaneous neoplasias.

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