Basal Cell Carcinoma Mimicking Dermatofibrosarcoma Protuberans: Case Report

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Abstract: ***Introduction:*** Basal cell carcinoma (BCC) is the most common type of carcinoma worldwide. BCC development is the result of a complex interaction between environmental, phenotypic and genetic factors. Presentation is highly heterogeneous, presenting from superficial or nodular lesions with a good prognosis to very extensive lesions in atypical locations clinically and radiologically mimicking other tumors. ***Case presentation:*** We present the case of a 34-year-old man with no past medical history, with an asymptomatic nodule that was increasing in size on the right thigh. High-frequency ultrasonography (HFUS) evaluation revealed an oval-shaped lesion with mixed echogenicity and posterior enhancement, with Doppler showing increased vascularization. A biopsy of the lesion was performed with suspicion of dermatofibrosarcoma protuberans, reporting unexpectedly an infiltrative basal cell carcinoma. The tumor was excised with free margins in the final pathology report. ***Conclusions:*** HFUS is an increasingly utilized tool that can accompany the study of a malignant lesion. Further studies are required to define more specific criteria for different skin tumors, always in conjunction with histology.

Keywords: Basal Cell Carcinoma, Carcinoma, Dermatofibrosarcoma Protuberans, High-Frequency Ultrasonography, Pathology.

INTRODUCTION

Basal cell carcinoma (BCC) is a malignant epithelial tumor that originates from pluripotent stem cells located at the level of the hair follicles and the interfollicular epidermis (1). The likelihood of developing a BCC depends on multiple genetic, phenotypic, and environmental factors. In relation to its genetic predisposition, mutations in the "Sonic Hedgehog" signaling pathway have been implicated, mainly in the PTCH1 gene (patched 1), as well as mutations in the p53 tumor suppressor gene, isolated in up to 44-65% of tumors (1). Regarding the phenotypic characteristics, various risk factors have been reported, such as family or personal history of skin cancer, light phototypes (I, II and III) and age (1,2). Of the environmental factors, the one that has a significant impact on the development of BCC is intermittent exposure to ultraviolet radiation, mainly in childhood and adolescence. (1)

Although the incidence increases considerably with age, with a peak of presentation between 80 and 89 years of age, in recent decades there has been an increase in the presentation of BCC at younger ages, currently being 2 times more frequent in women of the age range between 40 and 49 years than in previous years (3). Most cases of BCC occur in the head and neck, followed by the trunk and extremities (3,4). Mortality associated with BCC is extremely low, with reports ranging from 0.0028% to 0.55%; however, approximately 20% of BCCs have local aggressive and destructive behavior, which negatively impacts the quality of life of patients (3)

The clinical presentation of BCC is quite variable, from a dome-shaped pearly euchromic papule to rapidly evolving destructive infiltrative nodules or lesions that may invade deep anatomic structures. (5) The diagnosis is made by biopsy, which will indicate the histological subtype, the level of tissue invasion and the presence or absence of perineural involvement, in order to define the behavior. (6). Depending on the clinical and histopathological subtype, there are multiple inflammatory and tumoral entities in the group of differential diagnoses of BCC and that must be taken into account as differential diagnoses or large "mimetics". Among these lichenoid keratoses, amelanotic melanoma, Merkel cell carcinoma , trichoepithelioma and dermatofibrosarcoma protuberans. The latter, is a rare, clinically indolent, slow-growing dermal sarcoma with a high rate of local recurrence and infiltration, but low metastatic potential. (7) Surgery is the standard treatment for most BCCs. Standard excision or micrographic surgery (Mohs) can be used depending on the characteristics of the tumor (size, location, previous recurrences, histology). Mohs micrographic surgery is reserved for high-risk tumors, recurrent BCC, or BCC in highly functional anatomic sites. (8).

Here we present the case of a 34-year-old man with an asymptomatic nodule that was increasing in size on the right thigh. High-frequency ultrasound and clinical findings were consistent with dermatofibrosarcoma protuberans, however, biopsy unexpectedly reported infiltrating basal cell carcinoma. The tumor was removed with free margins in the final pathology report.

CASE REPORT

A 34 year old male with no past medical history, presented with an enlarging asymptomatic nodule on the right thigh Fig 1A. A high frequency ultrasonography was performed showing an oval shape lesion with mixed echogenicity and posterior enhancement (Fig 2 A-B), with power doppler revealing prominent vascularity of

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the lesion. With these findings an initial clinical diagnosis of dermatofibrosarcoma protuberans was made. Subsequently, a biopsy was made, confirming the diagnosis of infiltrative basal cell carcinoma exhibiting classic trabecular cords of few basaloid keratinocytes in a myxoid stroma. The tumor was excised with free margins on the final report of pathology.

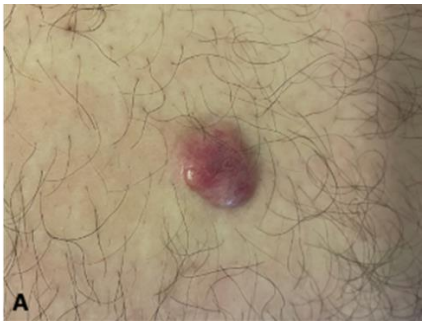

Figure 1: In the right thigh we observed an erythematous-violaceous nodule with a smooth surface, mobile, not adhered to deep planes of 3.8 x 3.5 cm, findings suggestive of dermatofibrosarcoma protuberans..

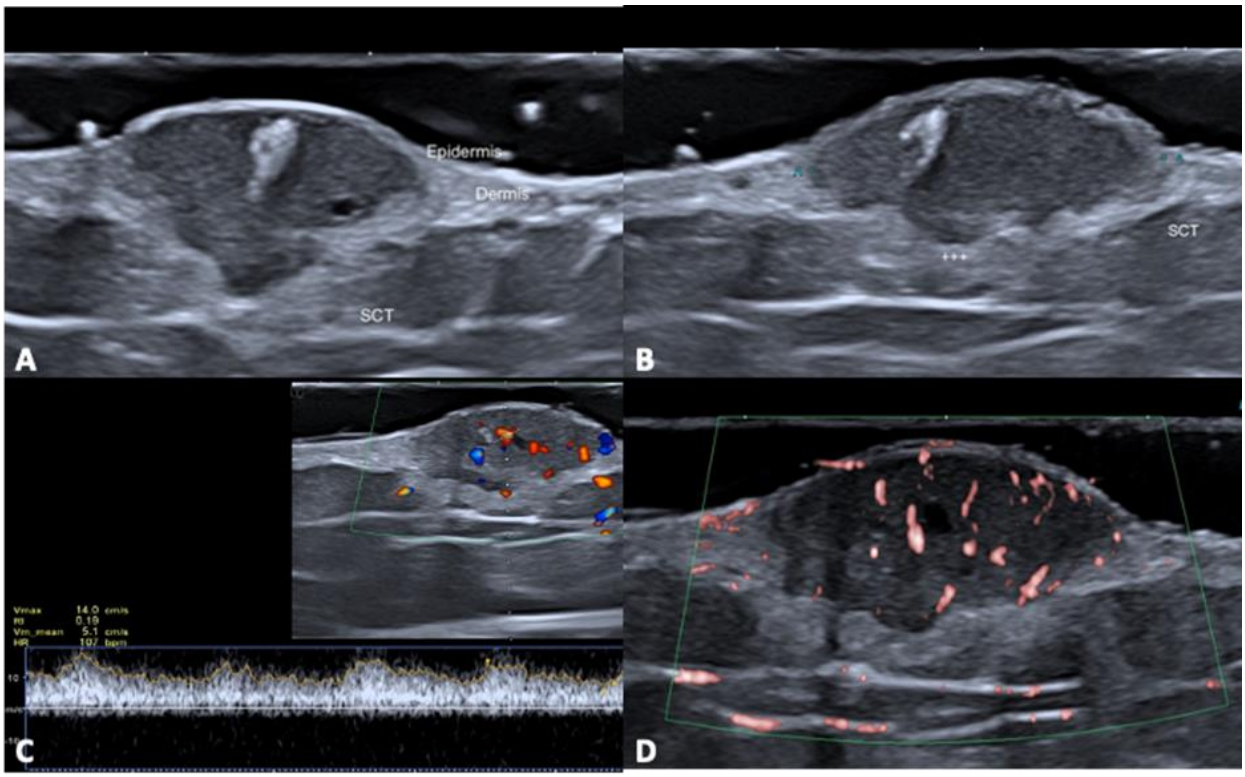

Fig 2. A, B Grey scale ultrasound axial and longitudinal view shows a oval-shape, with mixed echogenicity, dermal and hypodermal mass. Note the lower echogenicity in the upper part and the hyper-echogenicity at the deeper part (+++) , the lobulated and convex borders at the deep part of the tumor. **C,** Power Doppler longitudinal view demonstrates prominent vascularity inside the lesion. **D,** Color Doppler Duplex ,longitudinal view, demonstrates arterial vessels with low resistant flow .

DISCUSSION

Basal cell carcinoma, a subtype of non-melanoma skin cancer, is the most common malignancy worldwide characterized by its low metastatic potential, with an increasing incidence (2, 4,9). Caucasians are the most common affected population, due to its association with fair skin and UV radiation (10). Other risk factors include family history of skin cancer, living in rural areas, personal history of more than 10 sun burns and actinic conjunctivitis as reported by Sanchez, et al in Colombia (11); as well as a higher prevalence increasing with age and in males. All of this underlines the importance of the environmental, phenotypic and genetic factors (8).

Clinically presentation varies, however it usually arises in a sun-damaged skin as a non-healing lesion, with classical characteristics including ulceration, pigmentation and telangiectasias (4,12). Nodular basal cell carcinoma is the most common clinical subtype, presenting as a pearly papule/ nodule with arborizing vessels on the head and neck. Other clinical presentations include superficial, most commonly located on the trunk as fibroepithelial subtype, and morpheaform and infiltrative located on the head and neck (9). However, these lesions can be clinically misdiagnosed with other benign and malignant entities, especially when located on the lower limbs, given its uncommon anatomic presentation site. There is few literature of BCC on the lower limbs. Hooshang, et al reported a prevalence of lower limb BCC of 1.53% with

a mean age of 69 years, male predominance, and nodular as the most common pathological subtype (13); while Pranteda et al (14), Suppa et al (15) and Wolner (16) have reported prevalence of 7, 7.9% and 13.8% respectively. On the other hand, Pearson, et al described in a 150 series of BCC on the lower extremities that there was a female predominance, with the superficial subtype as the most common, and most frequently arising below the knee (17). Furthermore, it has been noted by Lombardi et al that less than half of BCC in this location displayed classic BCC dermoscopy criteria, with squamous cell carcinoma being the main differential diagnosis; and erosions/ulcerations and polymorphic vessels concerning features for malignant-looking BCC (9).

As shown in the present case, dermatofibrosarcoma protuberans (DFSP) can be a mimicker of BCC in the lower extremities. DFSP usually presents as a slowly progressive lesion on the proximal extremities or trunk (18). On ultrasound, an imaging study useful both for diagnosis and for predicting the degree of subclinical extension of the lesion (19) it usually appears as an oval, hypoechoic or mixed echogenic lesion located in dermis and subcutis, finger-like projections at the base of the tumor, lobulated margins and hyperechoic enhancement of the surronding tissues are very characteristic of this tumor (19, 20). The findings of oval lesion, lobulated margins, and posterior enhancement, as seen in this case, are sugestive findings of DFSP,so for that the presumtive diagnosis by ultrasound was DFSP. Moreover, Doppler activity shows important increased vascularity , a finding that is less common in BCC(21). Regarding BCC, it usually appears as a hypoechoic dermal or subdermal lesion with variable shape containing classic hyperechoic spots (22) (Fig 3).These spots have bee associated with compact nest of neoplastic cells, and are considering a predictor of aggressiveness of the BCC(23).At the ultrasound exploration our patient do not have hiperechoic dots inside the lession. The absence of hyperechoic points in BBC has been reported but in very small superficial tumors, however in BCC with the size observed in this case they are generally present. It is noteworthy that the ultrasonographic evaluation of our patient was carried out with high-resolution 18 MHz ultrasound that allows the visualization of these points and by a Radiologist with more than 10 years of training in dermatological ultrasound that guarantees the proper performance of the study.Another interesting point is that the vascularization of BBC at the Doppler exploration usually is low to moderate , differing with what was found in the high frequency ultrasonography (HFUS) of this case. So, even though HFUS is becoming a very significant toll due to its accessibility and non-invasive nature, still does not replace pathologic study for the final diagnosis.

This case shows an atypical presentation of BCC with trabecular growth pattern on histology, localized on an uncommon site. The latter emphasizes the various clinical forms of BCC which can be easily misdiagnosed by BCC mimicking lesions even with the support of other diagnostic modalities such as HFUS. For that reason, histopathology confirmation is mandatory in lesions suggesting malignancy.

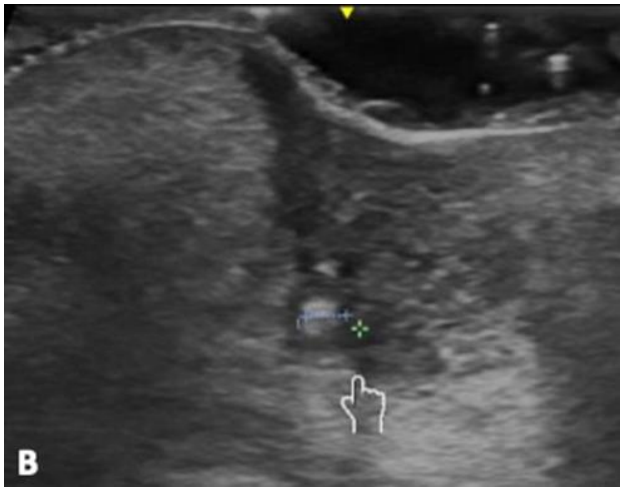

Figure 3: Grey scale ultrasound axial view shows the classic ultrasound aspect of basal cell tumor with hypoechoic, irregular and ill defined nodule; the marker shows the typical hyperechoic dots inside it.

CONCLUSIONS

The differential diagnosis of BCC is wide, and includes DFSP as the case presented. Furthermore, the atypical presentations of BCC in lower extremities is more common, in which the clinical differentiation of BCC from mimicking lesions is not well established.

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