Facial extensive ulcer

Úlcera extensa facial

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Article received: 1/30/2015
Accepted for publication: 2/17/2015

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http://dx.doi.org/10.1590/1806-9282.62.01.21

Summary

Basosquamous carcinoma (BSC), as described in 1910, is a distinctive variety of skin cancer and its etiology and pathological characteristics have generated much controversy over the years. Currently, BSC is considered a basal cell carcinoma (BCC) subtype with aggressive behavior and greater tendency for recurrence and metastases. We present a clinical case recently reported in our unit.

Keywords: skin neoplasms, skin ulcer.

Basosquamous carcinoma (BSC), as described in 1910, is a distinctive variety of skin cancer and its etiology and pathological characteristics have generated much controversy over the years.[1] Currently, BSC is considered a basal cell carcinoma (BCC) subtype with aggressive behavior and greater tendency for recurrence and metastases.

Case

A 52-year-old male presented with a 6-year history of an impressive extensive ulcer on his right cheek (Figure 1). It appeared over the old scar of a previous BCC excised ten years ago. It had painless steady increase in size with progressive difficulty in ocular movements and epiphora. A facial CT scan demonstrated a destruction of the right maxilla and nasal bone with no lymphadenopathies in the head and neck regions. Blood cell counts, urea, sedimentation rate, electrolytes, urinalysis tests were within reference ranges and antinuclear and anti-neutrophil cytoplasmic antibodies (ANCA) were negative. Mycological and bacterial cultures were also negative. A skin biopsy was performed and sent for hematoxylin-eosin and immunohistological staining (Figure 2).

Discussion

The incidence of BSC is between 1.5-2.7% in the largest studies reported.[2] Local aggressive growth pattern and high potential for distant metastases are the main concerning facts. The rate of local recurrence of the BSC is 45%, almost twice as much as squamous cell carcinoma (SCC) and BCC.[3] This rare skin carcinoma has a metastatic rate of 5-8.4%. Prognostic factors for recurrence include positive resection margins, lymphatic invasion, perineural invasion and male gender.[4]

Clinical image of the lesion is non-specific but long-standing time of evolution is a common feature in all the patients reported. The majority of these tumors arise on the head and the neck (80%), the central face and perinasal areas being the most prevalent locations.[5]

Histological examination exhibits findings of both BCC and SCC with a transition zone. It is not clear if BSC develops *de novo* or evolves from a pre-existing lesion, but the "squamatization theory" is the most commonly accepted. Unfortunately, the patient's previous BCC that was excised 10 years before could not be re-evaluated. Immunohistochemical stains have helped to a better characterization of BSC. Areas of BCC are Ber-EP4, AE1 and AE3 positive. In contrast, CAM5.2 and a variable positivization of epithelial membrane antigen are identified in the SCC areas. Ber-EP4 stain is gradually negative in the transition zone.[6]

Non-prospective trials to compare the different therapeutic regimes to approach BSC are available. There are

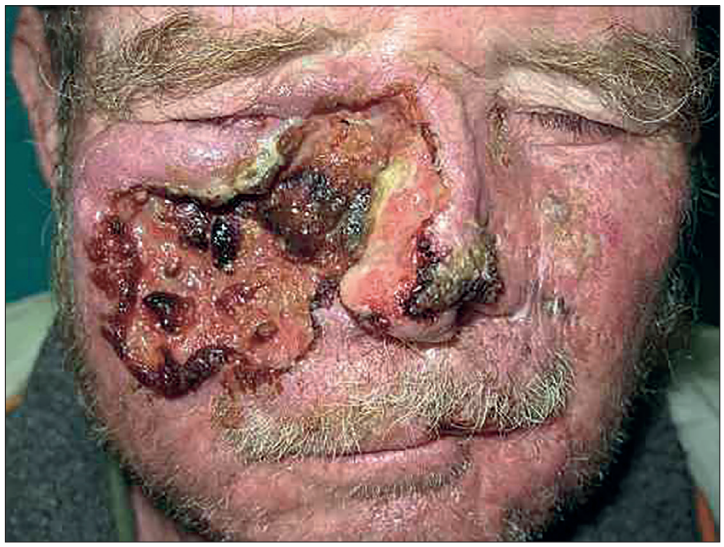

FIGURE 1 A 6-year slow growing facial ulcer developed over an old scar.

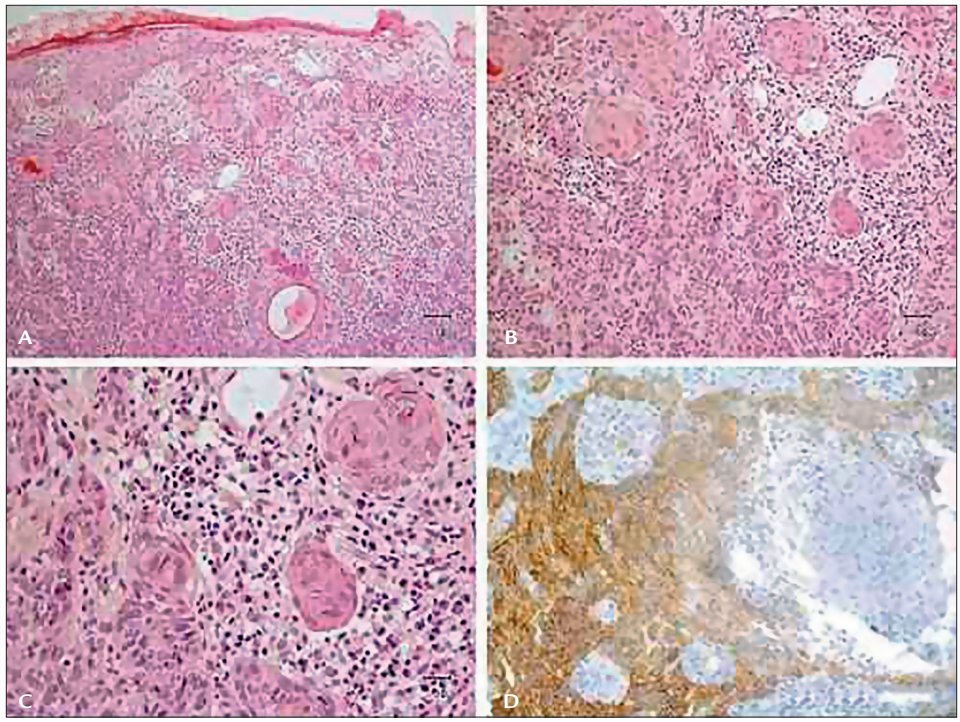

FIGURE 2 Infiltrative basal cell carcinoma (left side) adjacent to a squamous differentiation area with neoplastic cells containing eosinophilic cytoplasm and central keratinization (right side) (A,B,C). Positive immunostaining with Ber-EP4 in basaloid areas, contrasting negative Ber-EP4 in squamous differentiation area (D).

also no specific consensus management guidelines reported in the literature. Due to its aggressive local behavior pattern, Mohs micrographic surgery is recommended and should be standardized as it occurs in other skin tumors such as dermatofibrosarcoma protuberans.[7] Local radiation is a fitting alternative in selected cases. Curettage and fulguration or cryotherapy should not be considered unless the patient refuses an invasive surgical procedure. Vismodegib is the first selective inhibitor of the Hedgehog signaling pathway to be approved for the treatment of locally advanced and metastatic BCC. It could add a different therapeutic approach in advanced BSC; however, there is no case reported to date. The introduction of vismodegib, an orally active inhibitor of the Hedgehog signaling pathway by binding to Smoothened, approved for the treatment of locally advanced and metastatic BCC

could add a different therapeutic approach in advanced BSC. There is no case reported to date.

In patients with localized disease, metastasis to the regional lymph nodes is the strongest predictor of recurrence and survival. Selective sentinel node biopsy should be considered even in absence of palpable lymphadenopathies in order to manage a proper staging of this entity.[8]

In conclusion, we suggest that all dermatologists should be aware that BSC may not be focused in the same terms as BCC and SCC. Individualized guidelines should be employed.

Resumo

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Carcinoma basoescamoso, conforme descrito em 1910, é uma variedade distinta de câncer de pele e sua etiologia e características patológicas têm gerado muita controvérsia ao longo dos anos. Atualmente, BSC é considerado um subtipo de carcinoma de células basais com comportamento agressivo e maior tendência para a recorrência e metástase. Nós apresentamos um caso clínico relatado recentemente em nossa unidade.

Palavras-chave: dermatopatias, neoplasias cutâneas.

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