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Basal cell carcinoma of the scalp x frontal fibrosing alopecia: diagnostic and therapeutic challenge

Carcinoma basocelular de couro cabeludo x alopecia frontal fibrosante: desafio diagnóstico e terapêutico

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ABSTRACT: BCC is considered the most common neoplasia in the world, it can appear throughout the body including the scalp. Frontal fibrosing alopecia is a primary scarring alopecia, variant of lichen planopilaris. The association between the two pathologies has not been previously reported in the literature. In this case it is presented a brazilian female patient complaining of hair loss, with histopathological diagnosis of AFF and scalp BCC. The distinction of tumor margins for neoplastic excision is complex only by dermoscopy and physical examination, due to areas of common atrophy. So Mohs micrographic surgery was chosen for histopathological delimitation of margins.

Keywords: Fibrosing frontal alopecia; Basal cell carcinoma; Scarring alopecia; Mohs micrographic surgery.

RESUMO: O carcinoma basocelular é considerado a neoplasia mais comum do mundo, tem como principal fator de risco a radiação ultravioleta, pode aparecer em todo o corpo incluindo couro cabeludo. A alopecia frontal fibrosante é uma alopecia cicatricial primária, variante do líquen planopilar. A associação entre as duas patologias não tem relato prévio na literatura. Neste caso apresenta-se paciente feminina, pós-menopausa, atendida por queixa de queda de cabelo, com diagnóstico histopatológico de alopecia frontal fibrosante e carcinoma basocelular de couro cabeludo. A distinção das margens tumorais para exérese completa da neoplasia é complexa apenas pela dermatoscopia e exame físico, devido á presença de áreas de atrofia em comum. Então se optou pela cirurgia micrográfica de Mohs para delimitação histopatológica de margens.

Palavras-chave: Alopecia frontal fibrosante; Carcinoma basocelular; Alopecia cicatricial; Cirurgia micrográfica de Mohs.

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INTRODUCTION

Non-melanoma skin cancer (NMSC) correspond to 30% of all malignant tumors registered in Brazil. Basal cell carcinoma (BCC) and squamous cell carcinoma (SCC) are responsible for 99% of NMSC, with BCC three to five times more common than SCC[1]. Frontal fibrosing alopecia (FFA) is classified as primary scarring alopecia, histologically presenting inflammatory infiltrate around the hair follicles of probable immune cause, which places it as a variant of lichen planopilaris (LP). It was first described in 1994 and since then an increasing number of cases have been reported worldwide[2]. The purpose of this report is to expose the association between these two pathologies since we have not found any other description of a similar case. As well as extolling the benefits of Mohs micrographic surgery (MMS) in the treatment of cases in which the demarcation of margins is difficult by dermatoscopy and in which the preservation of viable skin is essential for the good functional and aesthetic result.

CASE REPORT

Female patient, 55 years old, white, menopause at 47 years old. She started monitoring in a dermatological office due to hair loss that started after gastroplasty, with progressive worsening, even with the use of a compound with proteins and nutrients that favor hair growth, prescribed by the digestive tract surgeon. In addition to areas of frontal alopecia with atrophic skin, the patient had areas of linear alopecia that entered the scalp, bordering the midline of the hair. The first diagnostic hypotheses were Chronic Cutaneous Lupus Erythematosus, Saber-scleroderma or marginal frontal alopecia, but the scalp biopsy clarified the diagnosis. Histopathological examination showed a pattern of scar alopecia compatible with FFA and scalp BCC. The patient was referred to the dermatological surgeon for excision of the lesion. In the consultation with this professional, a second suspected lesion of BCC was identified very close to the first, also in the area of alopecia. In view of the above, in surgical planning, Mohs micrographic surgery was chosen in order to preserve a larger area of viable skin and to define margins with greater accuracy. The surgical procedure was performed with total removal of the neoplastic lesions, without the need for enlarging the margins.

DISCUSSION

BCC is considered the most common malignancy in the world, more frequent in men, uncommon in high phototype, can occur at any age, but more than 75% of patients are over 40 years old. The main factor involved in the pathogenesis of BCC is UVR, in particular type B[1]. Although the prevalence of this tumor increases according to sun exposure, the distribution of the lesions does not correlate directly with the most exposed area, they can appear in several areas of the body, including the vulva and scalp, areas usually physically protected from UVR. A study that evaluated scalp BCC showed an incidence of 2.6% of injuries in this region[3]. The risk factors described are exposure to UVR, scalp radiotherapy and immunosuppression[4].

In the case of the patient in question, there is a report of excessive exposure to UVR without photoprotection during childhood, but at this time in life, there was still no report of alopecia, so the scalp was protected by the presence of hair. Although the lesions appeared in the area of alopecia, the time interval between the complaint of alopecia and the diagnosis of BCC was short, so the exposure of the scalp to UVR was not prolonged, which is usually necessary for the development of the tumor.

Initially described in Australia in 1994, FFA is a variant of LP, characterized by a progressive form of scarring alopecia with marginal frontotemporal and eyebrow rarefaction in female patients, especially after menopause. The diagnosis must be confirmed with histopathology[5]. The pathophysiology of the disease is not completely known, it is known that there is the formation of an immunomediated inflammatory infiltrate characterized by the prevalence of CD8 lymphocytes in the follicular bulb, leading to the permanent destruction of these follicles[6].

The presence of a chronic inflammatory reaction in the pathophysiology of FFA may be at the heart of the relationship with BCC. In pathology, peritumoral inflammatory infiltrate, predominantly lymphocyte, is frequently observed. This fact shows the importance of the immune system in destroying tumor cells. The concept of "immunologically vulnerable areas" would be related to the difficulty in circulating natural killer cells or to angiogenic stimulation, favoring the appearance of neoplasms[7]. However, as we did not find other reports with the same association, we cannot say that there is a causal factor between the two entities.

In dermatoscopy, both diseases have well-defined characteristics, but the presence of white areas due to fibrosis of the dermis is present in common, which makes it difficult to distinguish the tumor margins. In FFA, dermoscopy of the scalp reveals loss of follicular ostia openings, perifollicular erythema, perifollicular scales adhered to the base of capillary stems and white areas of fibrosis[8]. In BCC, dermoscopic structures are divided into vascular, pigmented and non-vascular / non-pigmented changes. Arboriform vessels are described as the main structures identified in BCC dermatoscopy, followed by telangiectasias. Among the pigmented structures, the most common are the blue-gray ovoid nests. Structures that are neither pigmented nor vascular can present as ulcerations, and reddish-white areas that correspond to areas of dermis

fibrosis[9].

In the patient in question, due to the difficulty of delimiting the lateral margins of cancerous lesions, as they are overlapping a scar area, treatment was indicated for Mohs micrographic surgery. This surgical technique was described in 1933, it differs from the conventional technique in that it allows the assessment of 100% of the tumor margins, and in sparing areas of healthy skin, reducing not only the size of the surgical wound, but also the chance of recurrence[10].

With the inspection of the scalp, it was possible to delimit the FFA area, with dermoscopy, tumor lesions were identified and a margin of approximately 3mm was marked (Figure 1). The surgical procedure started with tumor debulking, followed by margin resection (Figure 2). The freezing and cuts of the piece were performed by the surgeon himself in the microtome cryostat, with cuts of 10 microns thick, followed by fixation, staining and histopathological analysis of the slides by the same professional.

Figura 1: Demarcação de área de alopecia frontal fibrosante (AFF) e de lesões neoplásicas com margem de 3mm.

Figura 2: Ferida cirúrgica após debulking tumoral e ressecção de margens.

Histopathology allowed a clear differentiation between the two conditions, and the complete resection of the tumors, without the need for margin expansion. The BCC appears as basaloid tumor cells sprouting from the epidermis, from follicles or inside the dermis, with variable atypia. Often retraction artifact can be noticed and nuclei arranged in peripheral palisade. Around the tumor, infiltration of lymphocytes and plasmocytes varies[11]. In FFA, the findings include prominent perifollicular fibrosis, lymphocytic inflammation with lichenoid pattern around the infundibulum region, isthmus and follicle protuberance and reduction in the number of follicles with replacement by fibrous tracts.

This study illustrates an association not previously found in the literature that challenges the diagnosis of neoplasia in areas chronically affected by other dermatological diseases and the delimitation of margins for the complete removal of neoplastic lesions, without extensive resection of healthy skin. It also raises the hypothesis of a causal relationship between FFA and BCC and opens space for further reports involving this association.

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