

Metatypical BCC of The Ear: Partial Amputation of The Auricle as Adequate Dermatological Approach

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Non-melanoma skin cancer is the most common cancer in the world (1). Basal cell carcinoma (BCC) is the most common malignant neoplasia of the skin, accounting for approximately 80% of all cutaneous tumors (2). High cumulative ultraviolet (UV) light exposure and the presence of Fitzpatrick skin phototype I or II are pathogenetic risk factors (3). UV radiation induces mutations in the tumor suppressor PTCH1 gene, leading to increased Hedgehog (XX/Hh) signaling pathway activity (4). Dysfunction or abnormal activation of the Hedgehog signaling pathway is associated with disorders of embryonic development and the development of cancers such as Goltz–Gorlin syndrome for example (5).

BCC has the characteristics of a locally invasive, slow-growing skin cancer, with a metastatic incidence between 0.0028-0.5%, with areas in the head and neck most commonly affected (6).

Metatypical basal cell carcinoma (MTBCC) is a rare variant of BCC that has histopathological characteristics of both basal cell carcinoma (BCC) and squamous cell carcinoma (SCC) (7).

We describe a 53-year-old patient who presented to our dermatology clinic for the first time due to a slow-growing wound of the left ear with a four-year history. He reported a ten-year period, during which, during the summer trimester, he had travelled weekly to the seaside and back, and it was the left side of his face, including the auricle, that had been exposed to intense solar radiation.

According to his history, he had visited a skin doctor for the first time a year ago (Fig. 1a, 1b). Subsequently, or a year later, on clinical examination, an ulcero-necrotic lesion was found in the left ear region and a biopsy was taken. Histopathological verification was suggestive in favor of the diagnosis of metatypical basal cell carcinoma. The ulcerative changes were then approximately 1.5 cm in length and 1 cm in width of the helix, with infiltration in depth, disrupting the integrity of Darwin's tubercle (Figs. 1c, 1d).

At the time of admission, one year later, the disease changes involved the skin of the left ear and were represented by a dense

necrotic formation in the region of the left ear, with irregular borders and an uneven surface, covered in places with greenish-brownish crusts, measuring about 2.5cm in length and width (Figure. 1c, 1d). The tumor formation covers the helix and anthelix in depth, adjacent to the concha (Figures 1c,1d).

The patient denied having any concomitant diseases. He reported history of disc herniation surgery in 2012 and penicillin allergy.

Paraclinical studies were within reference limits. Screening showed no evidence of process dissemination. A partial resection of the auricle was performed (Figure 2a-2f). Subsequent histologic examination again verified evidence of metatypical basal cell carcinoma with clear resection lines, stage T2N0M0R0.

MTBCC was first described by MacCormac in 1910 as a histologic variant in a series of rodent ulcers where basal cell and squamous cell tumors were found within a single lesion as collisional tumors (8).

In 1928, Montgomery defined this morphology as a neoplasm completely distinct as a nosological entity (9). In 1974, the WHO confirmed the distinction of MTBCC from BCC and SCC (10). As a clinical manifestation, MTC resembles BCC, and this specificity makes it difficult to make an accurate diagnosis on the basis of clinic or dermatoscopy alone (11). Despite their similar macroscopic appearance, the two types of lesions have different clinical behavior (12). BCC metastasizes infrequently, whereas MTBCC is more similar (in terms of clinical behavior) to squamous cell carcinoma of the skin and has a generally worse prognosis (13).

Important factors for the prognosis of MTBCC are: sex, anatomical localization, histopathological variant of the tumor, tumor size, positive resection lines, lymphovascular and perineural infiltration with tumor cells (14),(15).

A number of retrospective studies of the head and neck BCC have shown a distinct predominance of males (65%) versus females (35%) (14). Specific sites on the face (nasolabial fold, nasal,



Figure 1 : a,b: Clinical status: ulcerative lesion with irregular shape affecting the helix of the left auricle measuring about 1x1,5cm.
Figure 1 : c,d: Clinical status: ulcero-necrotic lesion in the left auricle involving the helix and anthelix with approximate dimensions of about 2x2, 5cm

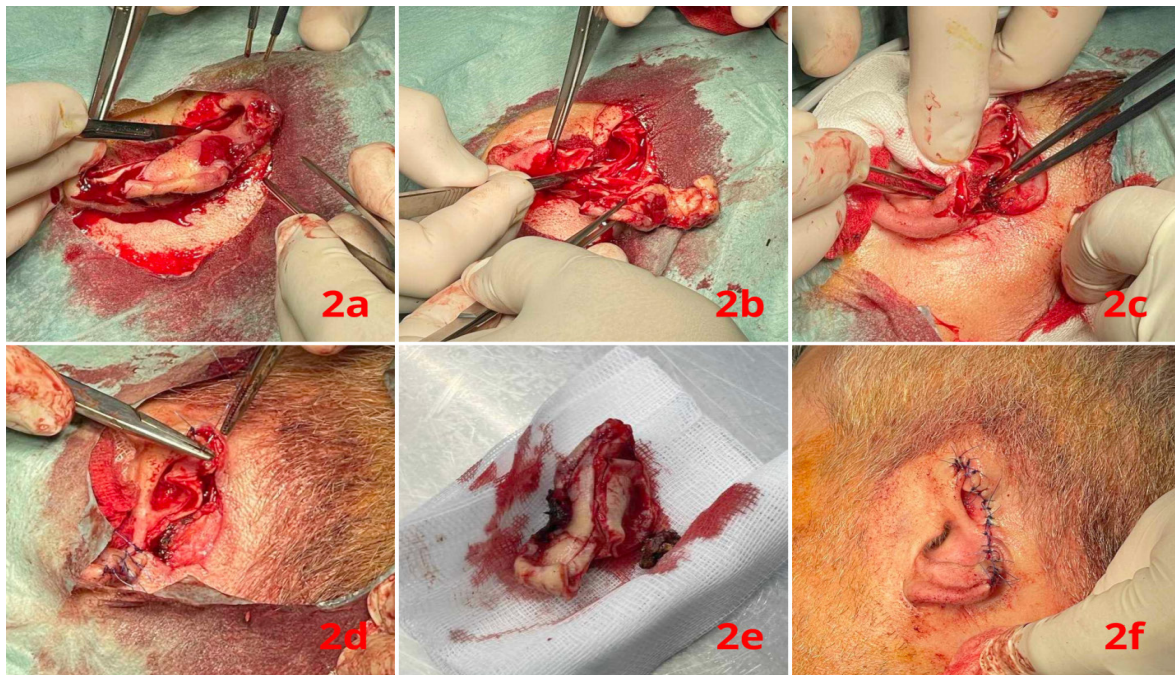


Figure 2 : a,b: Intraoperative photograph of partial resection of the ulceronecrotic lesion with an operative margin of 0.5 cm.
Figure 2c: Intraoperative photograph after partial resection of the ulceronecrotic lesion in the left auricle - cauterization a
Figure 2d: Intraoperative photograph after partial resection of the ulceronecrotic lesion in the region of the left auricle - staged suturing with single continuous sutures.
Figure 2f: Postoperative photograph of the removed tumor in the auricle area.
Figure 2d: Postoperative photograph immediately after cartilage resection, adpatation of the surrounding skin and closure of the defect

orbital, and auricular regions) have been described as high-risk areas ("H-zone") for the development of basal cell carcinoma because they are associated with an increased recurrence rate relative to other anatomic sites (16).

Ulcerative changes are more characteristic of aggressive histopathological types, which include metatypical basal cell carcinoma (14).

BCCs in the ear region are characterized by a higher risk of recurrence at tumor size (10-20 mm), whereas those in the neck and nose region recur at lesion size greater than 20 mm (17). Of note, the recurrence rate increases when the lesion size is ≥ 10 mm at the time of excision (17).

According to a study examining 7732 cases of basal cell carcinoma in the head and neck region, only 9% of the BCCs examined were in the ear region, and they were characterized by larger lesion size, required a greater number of Mohs re-excisions, and left a larger defect compared to BCCs in other anatomic regions (18). Separately, there is scientific evidence that basal cell carcinomas in the ear region are more likely to have an aggressive histologic phenotype (19).

Currently, the "gold standard" treatment for BCC is surgical removal, with a preference for Mohs surgery for high-risk types (20).

For primary, well-demarcated, low-risk BCCs less than 2 cm in size, a resection margin of 3 mm yields satisfactory results (21).

For tumors with high risk of recurrence and/or sizes greater than 2 cm, a surgical margin of 4-6 mm is suggested to obtain clear margins, with a minimum 5-mm resection margin recommended for high-risk tumors and 6-mm or Mohs surgery for recurrent tumors (21).

Currently, there are no clearly standardized criteria for the treatment of MTBCC in the literature, but wider resection fields of surgical certainty are recommended due to the high risk of dissemination (22).

Metatypical ear carcinoma is considered high-risk in terms of proliferation, dissemination, prognosis, and cosmetic outcome; therefore, radical removal of the tumor with a surgical margin of safety of at least 5 mm is necessary.

Modern medicine offers a solution for postoperative defects through the new possibilities of 3D planning and 3D printing of ear prostheses, which should improve the cosmetic outcome after radical surgery (23).

However, it should be emphasized that in cases with advanced stages of the disease, the primary goal is to achieve a long-lasting remission at the expense of the cosmetic outcome, i.e. the patient's health should be put first.

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