CASE REPORT

RJME
Romanian Journal of Morphology & Embryology
http://www.rjme.ro/

Giant basal cell carcinoma of the nasal pyramid – case report

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Abstract

Basal cell carcinoma (BCC) is a frequent form of skin cancer, which usually affects people that have been exposed to the sunlight for longer periods of time. The cells of the lower part of the epidermis are called the basal cell layer. These cells constantly divide to form new cells to replace the squamous cells that wear off the skin's surface. As these cells move up in the epidermis, they get flatter, eventually becoming squamous cells. Therefore, the BCC develops from these cells. Most BCCs have indolent behavior, with cure rates very high after low-complexity treatment. However, some lesions are very aggressive and there are only a few papers focusing on the subtype of this skin cancer known with the name *ulcus rodens* or giant BCC. In this study, we evaluate a case of *ulcus rodens* or giant BCC, subtype of the BCC skin cancer located in the area of the nasal pyramid, stage III, TxNxMx, with lymphatic and vascular invasion present.

Keywords: basal cell carcinoma, nasal pyramid, *ulcus rodens*, tumoral necrosis, inflammatory infiltrate.

Introduction

Skin cancer is one of the most common neoplasms. In 2018, around one million non-melanic cell cancers and 288 000 malignant melanomas have been reported globally [1]. According to some studies, the incidence of skin cancer increases exponentially throughout the world. In the US, in 2012 there were about 5.4 million non-melanic skin cancers. That is why it is believed that one in five Americans will develop skin cancer during their lifetime [2, 3].

In skin cancers, typically, the patients alert themselves over the cosmetic appearance and often address the medical services (dermatology or plastic surgery mostly) for this reason. The risk factors of this type of cancer are the increased rate of solar radiation exposure and the thinning or lack of ozone layer. The main etiological factor responsible for basal cell carcinoma (BCC) is the chronic ultraviolet (UV) radiation exposure at the expense mostly of UVB rays with 290–320 nm wavelengths [4]. This results in activating of proto-oncogenes and inactivation of tumor suppressive genes in the keratinocytes. Besides UV radiation, there are other exogenous carcinogens, such as exposure to the ionizing radiation, arsenic, industrial chemical substances, such as vinyl chloride, polycyclic aromatic hydrocarbons, as well as alkalizing agents [5–8]. In addition to environmental factors, genetic susceptibility and insufficient immunological surveillance are two constitutional factors involved in the etiopathogenesis of skin cancer. Cutaneous BCC is most often encountered in the white race and affects especially the skin of the head and neck segments of the body. Long-term exposures as well as short-term but frequent exposures to UV light could be major risk factors that lead to this type of cancer [9]. BCC is slow, but untreated can spread locally.

We present a case of giant BCC localized to the nasal pyramid that invaded the sinuses of the face and determined the lysis of the inner wall of the orbits.

Case presentation

Patient B.T., Caucasian male, 57-year-old, history of alcohol abuse, was admitted in the Ear, Nose & Throat (ENT) Ward, Emergency County Hospital, Constanţa, Romania, in June 2016, for nasal obstruction bilateral, bleeding from the nasal pyramid of unknown origin, view impairment, lack of smell, frontal headache, facial pain, bleeding and tumefaction, and infectious status (fever, sweating), symptoms and signs present for almost two months, excepting the fever, bleeding and sweating.

Clinical examination

Patient presents himself with a huge compressive bandage soaked in blood located above his nasal pyramid which, at the time of its removal, shows the presence of a massive tumoral lesion located in the middle of the face, where the nasal pyramid is normally found, and extending itself towards the internal angle of the orbit bilateral and clouding half of the view fields, with tissue destruction of the root of the nose, *dorsum nasi*, both nasal wings, columella, tip of the nose (Figures 1 and 2). The lower edge of the tumor was located at the upper lip level, while the upper rim ends between the eyebrows. At palpation, no bone structure is felt, just elastic tumoral mass.

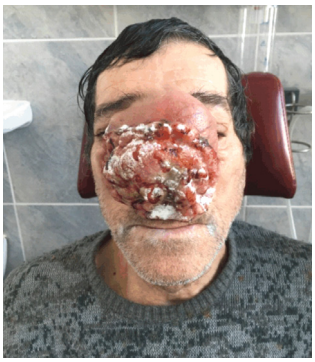

Figure 1 – ***Macroscopic image of the tumor. Front view.***

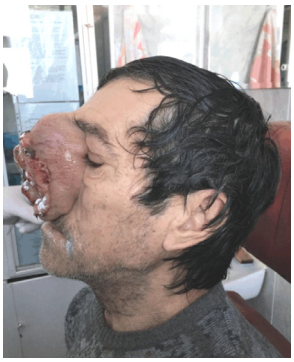

Figure 2 – ***Side view of the tumor at the level of the nasal pyramid.***

Anamnesis reveals that the giant lesion evolved from a smaller lesion, which resembled to a nodular elevated pearl-shaped lesion of the skin of the right side of the pyramid, and it subsequently extended during many years into an approx. 10 cm longitudinal and approx. 9 cm width tumor, with telangiectasia on the surface and periphery, and a big central ulcerative area with diffuse bleeding and necrotic spots spread inside the ulceration, and covered with little purulent yellowish secretions and hematic crusts and diffuse bleeding.

Imagistic exams

Computed tomography (CT) scan of the head and *viscerocranium* with contrast provides further information over the tumor extension: a tumor mass about 90/106/102 mm, irregular contour, with heterogeneous structure, and necrotic zones in the inferior and middle turbinates on the right side nasal cavity. The tumor formation was extended in the ipsilateral maxillary sinus, ethmoidal cells, and bilateral frontal sinuses, nasal septum and the partial invasion of the left ethmoidal cells (Figures 3 and 4).

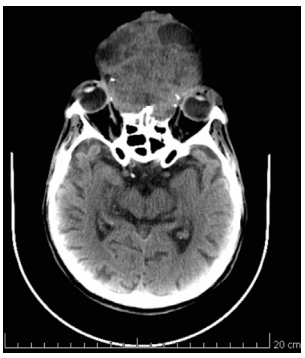

Figure 3 – ***CT image showing tumor enlargement to the frontal sinuses through the ethmoidal cells, especially anterior cells. Latent destruction of the internal wall of the orbits, not yet inside the orbit, thin layer of bone tissue still intact there and lack of nasal septum due to osteolysis.***

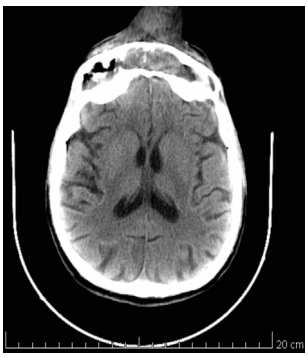

Figure 4 – ***The apex of the lesion outside the viscerocranium with the extension inside the frontal sinuses, with the preservation of the anterior wall of the frontal sinuses (due to its anatomical thicker structure).***

No nasal bones or cartilages of the nasal pyramid present in the outside-area occupied by the tumor. No intracranial invasion on CT scan.

Histopathological and immunohistochemical study

To determine the diagnosis, we considered it necessary to make some biopsies from the lesion. After local anesthesia, two tumor fragments of about 0.5/0.3/0.4 cm were harvested from ulcerated and bleeding areas that were placed in 10% formaldehyde solution. Then, the tumor fragments were included in histological paraffin, microtome-sized and stained with Hematoxylin–Eosin (HE). For the differential diagnosis, sections of 4 μm thick were made, which were plated with poly-L-lysine and prepared for immunohistochemistry studies.

The histopathological exam revealed the presence of a BCC with desmoplastic stroma, inflammatory infiltration, areas of tissue necrosis, with vascular tumoral invasion and without perineural invasion (Figures 5 and 6). Histopathological aspects were extremely varied from one area to another because of tumor size and ulcerated and infected areas. Tumor cells have a mixed, nodular and infiltrative growth pattern and the peritumoral inflammatory response was extremely varied as a quantitative expression.

For the immunohistochemical (IHC) study, we used the following antibodies: anti-cytokeratin (CK) 34βE12 (monoclonal mouse anti-human 34βE12, 1/50 dilution, Dako); anti-B-cell lymphoma 2 (BCL2) (monoclonal mouse anti-human BCL2, 1/50 dilution, Dako); anti-Ki67 (monoclonal mouse anti-human Ki67, clone MIB-1, 1/50 dilution, Dako); anti-cluster of differentiation (CD) 3 (monoclonal mouse anti-human CD3, clone F7.2.38, 1/50 dilution, Dako); anti-CD20 (monoclonal mouse anti-human CD20cy, clone L26, 1/50 dilution, Dako).

The IHC results of this study revealed an over-expression of the CK 34βE12 biomarker, with diffuse cytoplasmic staining in tumoral cells (Figure 7). Also, immunostaining for BCL2 showed an diffuse nuclear staining in tumoral cells (Figure 8). Evaluation of proliferative activity of tumor cells was done by using the anti-Ki67 antibody. In our case, the cell proliferation index was very low, less than 3% of the tumor cells present positive reaction at anti-Ki67 antibody (Figure 9). Regarding the inflammatory reaction, we found that it had a high intensity in areas of tissue necrosis. Peritumoral, the inflammatory reaction had a moderate intensity, with both T- and B-lymphocytes in variable amounts (Figures 10 and 11).

Further blood tests were performed. There was found a leukocytosis with neutrophilia (due to infection undergoing on the tumoral lesion), severe hypochromic microcytic anemia (hemoglobin 4.2 g/dL) due to constant diffuse bleeding from the lesion. Values of C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) were moderately increased (due to proliferative facial lesion), while other tests (coagulation tests, creatinine, urea, hepatic enzymes, bilirubin) had normal values.

The patient was treated for infectious status with antibiotherapy with large spectrum (Ceftazidime, 2nd generation cephalosporin, 1 g every 12 hours for five days), Cortisone, blood transfusion (Erythrocyte Mass and Plasma) and hemostatics to dissolve the hemoragic shock, opioid pain-killers, hydroelectrolytic rebalancing with 5% Glucose and 0.9% NaCl and local mild-compressive bandage each day after softly-cleansing the lesion with Iodine from the pus and Neomycin and Bacitracine mix-powder pulverization over the ulcerated area.

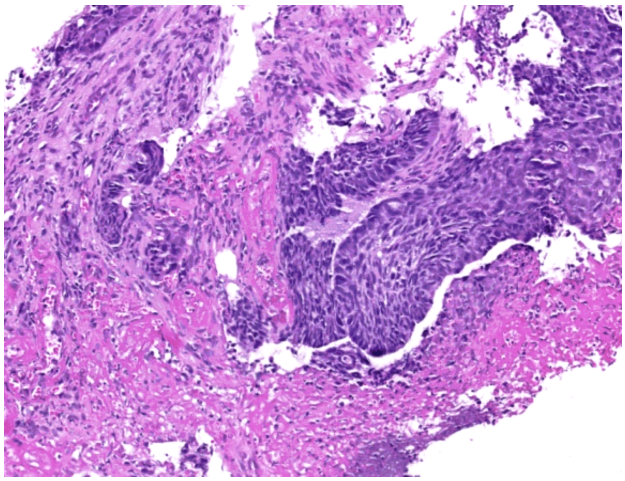

Figure 5 – ***General microscopic image of the tumor showing the nodular arrangement of the tumor cells, and the desmoplastic aspect of the stroma (HE staining, ×100).***

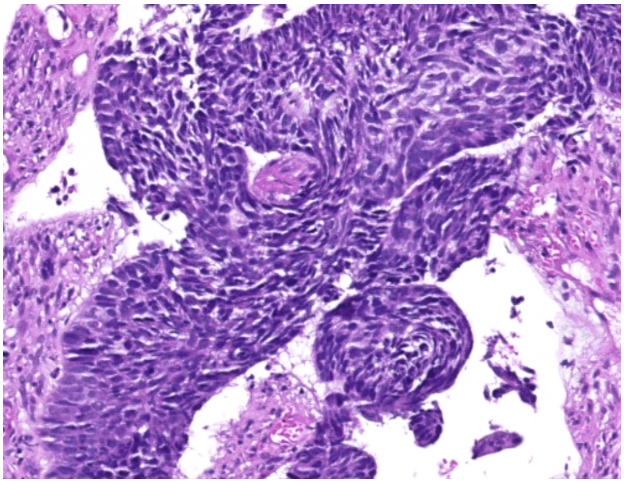

Figure 6 – ***Histopathological detail showing the appearance and layout of tumor cells (HE staining, ×200).***

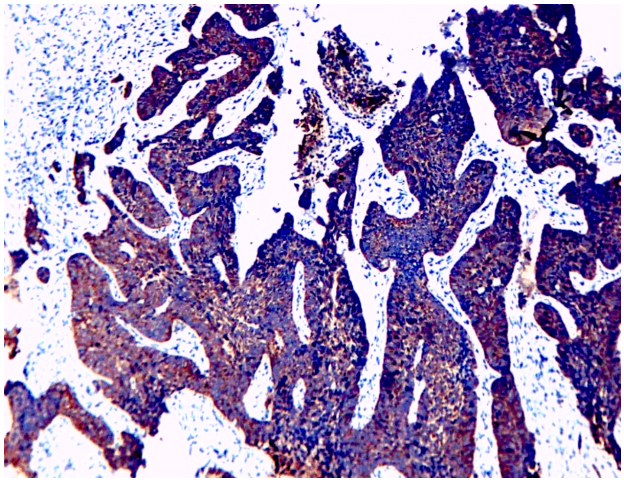

Figure 7 – ***Highly positive CK 34βE12 expression in cytoplasmic tumor cells (Immunostaining with anti-CK 34βE12 antibody, ×100). CK: Cytokeratin.***

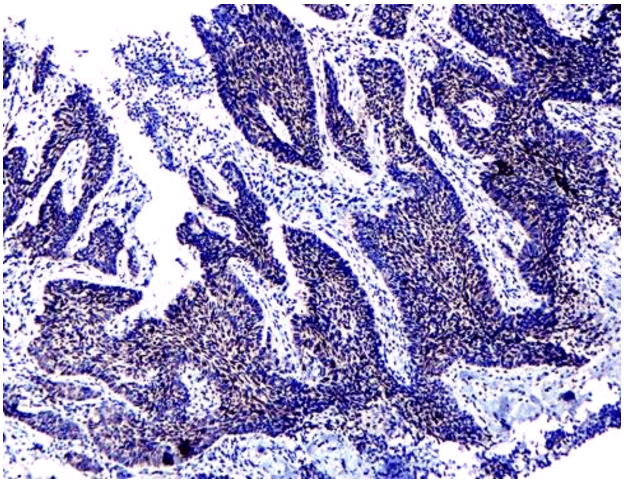

Figure 8 – ***Tumor cells with intense reaction to the anti-BCL2 antibody (Immunostaining with anti-BCL2 antibody, ×100). BCL2: B-cell lymphoma 2.***

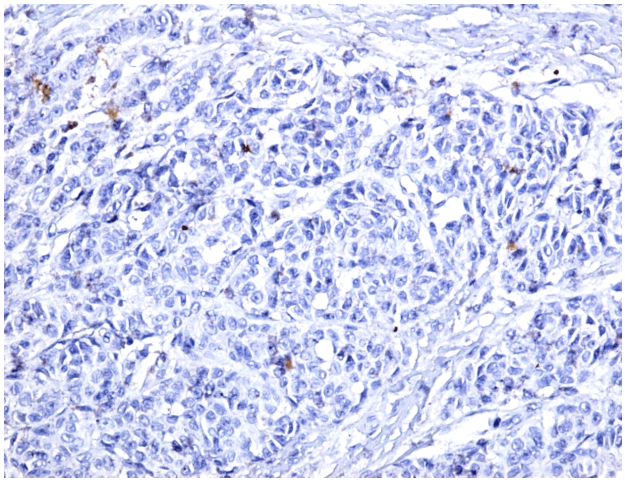

Figure 9 – ***Tumor cells with a very low proliferation index (less than 3%) (Immunostaining with anti-Ki67 antibody, ×200).***

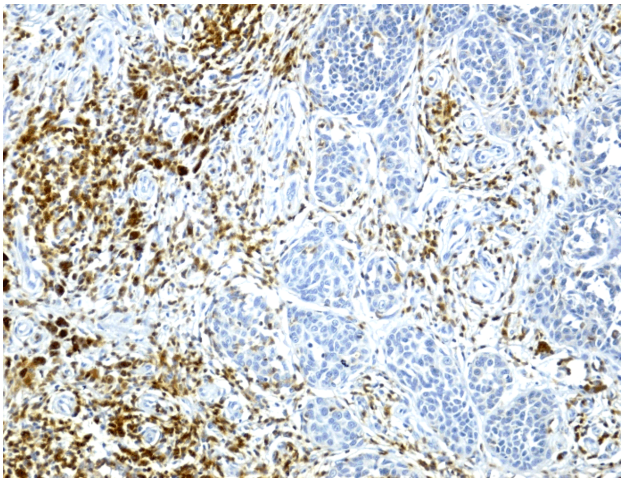

Figure 10 – ***Moderately peritumoral inflammatory infiltration with multiple T-lymphocytes dispersed in the stroma (Immunostaining with anti-CD3 antibody, ×100). CD3: Cluster of differentiation 3.***

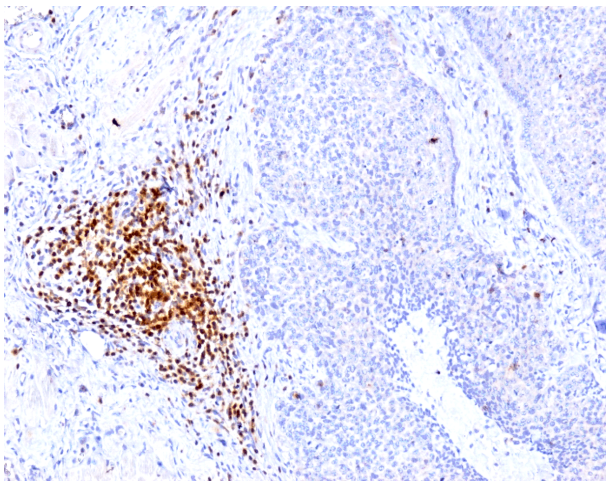

Figure 11 – ***Tumor stroma with low B-lymphocyte count (Immunostaining with anti-CD20 antibody, ×100). CD20: Cluster of differentiation 20.***

The patient was released from the Ward with the histological result in processing status, since it takes 3–4 weeks to be processed, but clinically stable with recommendation to claim the result of the histology after this period of time. The Hospital records show another admission in the ENT Ward, in June 2017, for re-balancing. ENT recommendations where to present himself in Oncology Ward for further therapeutic measures, until then he should avoid smoking, alcohol consumption, polluted atmosphere, local trauma to not increase the chances of massive bleeding, avoid the physical effort or other bleed-inducing actions in the area. Daily hygiene of the lesion with Iodine and compressive bandage at home or ENT Ward under medical attention.

Discussions

Here, we presented the case of a patient diagnosed with giant face tumor, a tumor that, according to histo-pathology and immunohistochemistry, proved to be an IIIrd stage BCC. The clinical aspect of the tumor suggested that this lesion progressed for a long time, being neglected by the patient.

The *BCC*, together with the *squamous cell carcinoma* (*SCC*), represent about 99% of non-melanic skin cancers [10, 11]. Of these two main forms of skin cancers, BCC represents about 75%, while SCC stands for 25% [12–14]. As such, we may state that the most frequent form of skin cancer is BCC.

Non-melanic cancers of the skin mainly affect white persons, aged over 65 years old [15–17]. In the last decades, there was observed an increase in the incidence of all forms of skin cancers, probably caused by a prolonged exposure to sun radiations [18, 19].

Most studies showed that BCC presents a low degree of malignity, low recurrence and a limited potential of metastasizing, but it also has the capacity of local invasion and tissue destruction [14, 17, 20].

Our presented case is classified as stage III giant BCC or *ulcus rodens*, TxNxMx, with lympatic and vascular invasion present without an endocranial invasion, regarding the fact the patient was not head-to-toe CT-scanned nor was he clinically presenting other metastasized areas, otherwise he would've been classified as stage IV. Even though BCC appears frequently, metastasis of the lymph nodes occurs very rarely, with a 0.1% rate [21]. There is a large variety of morphological skin appearances, such as micronodular, odular, infiltrative, superficial, sclerosing, morpheaform, keratotic cystic and pigmented variants [22]. Only 1% of BCC can lead to a giant BCC, because of the late addressability of most of the patients.

The treatment could be surgical in case of operability and medical in case of inoperability associated with metastatic events. There are only a few reports on the efficiency of the chemotherapy. In the case of systemic therapy, the following could be used: Methotrexate, Bleomycin, Vincristine, 5-Fluorouracil (5-FU), Cyclo-phosphamide, Dactinomycin and Toyomycin, Platinum and Taxanes [23]. In case of a surgical failure, Vismodegib, a Hedgehog pathway inhibitor could be used in a complicated, recurred form that has not responded to radiation [24].

Conclusions

Slow growth and poor prognosis are the main two characteristics of the BCC. With an evolving rate of just 1% into the giant form, precocious addressability could avoid a massive evolution of this form of skin cancer and could also improve the prognosis.

Conflict of interests

The authors declare that they have no conflict of interests.

Authors' contribution

Dănuț Dincă and Loredana Elena Stoica equally contributed to the manuscript.

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Received: November 30, 2018

Accepted: June 21, 2019