Giant Basal Cell Carcinoma of the Lumbosacral Region: A Case Report and Literature Review

Arzu Akçal¹, Selami Serhat Şirvan², Kamuran Zeynep Sevim³, Semra Hacıkerim Karşıdağ³, Kemal Uğurlu⁴
¹Department of Plastic, Reconstructive and Aesthetic Surgery, Akdeniz University School of Medicine, Antalya, Turkey
²Clinic of Plastic, Reconstructive and Aesthetic Surgery, Selahattin Cizrelioğlu State Hospital, Şırnak, Turkey
³Clinic of Plastic, Reconstructive and Aesthetic Surgery, Şişli Hamidiye Etfal Training and Research Hospital, İstanbul, Turkey
⁴Clinic of Plastic, Reconstructive and Aesthetic Surgery, Private Medistate Hospital, İstanbul, Turkey

Abstract

Objective: Although frequently regarded as a low grade malignancy, basal cell carcinoma (BCC) sometimes shows aggressive behavior. Giant BCC is defined as a lesion greater then 5 centimeter.

Material and Methods: The authors described the diagnosis and treatment protocol of giant BCC, involving lumbosacral region without any local or distant metastasis. Also the authors went through a thorough a retrospective literature research of giant BCCs in terms of frequency and treatment.

Results: Giant BCCs are biologically more aggressive; however, patient’s negligence is a key factor for the tumor size. International medical database (PubMed) search reveals 253 giant BCCs with the most commonly seen site as the face and back area. However perianal BCCs are seen lower then 1%.

Conclusion: In general, diagnosed giant BCCs account for lower then 1% of all BCCs. One must keep in mind this diagnosis to discard the metastasis rate among BCCs.

Keywords: Skin cancer, giant basal cell carcinoma, BCC

INTRODUCTION

Basal cell carcinoma (BCC) is the most prevalent malignancy among humans, and accounts for 75% of diagnosed skin cancer.¹ It is observed to be more common among Caucasian males. Since its primary cause is constant exposure to the sun, it is more frequently encountered in the head and neck region. BCC incidence increases by 3% globally every year.²

Although basal cell carcinoma is considered to be a low malignancy-grade tumor, it can occasionally display an aggressive character. The American Joint Committee on Cancer (AJCC) staging system has defined BCC’s larger than 5 cm in diameter as giant BCCs.³ The first personal experience with Giant BCC (GBCC) was reported by Eckhoff³ in 1951. GBCCs are rare, metastatic tumors that can demonstrate aggressive biological behavior and deep tissue invasion. Similar to smaller BCCs, GBCCs mostly arise from the basal layer and the adnexal cells of the epidermis in persons who are constantly exposed to the sun. There are a limited number of publications on GBCC in the literature.

MATERIAL AND METHODS

A search was conducted in PubMed on articles published from 1951 to 2014 using the keywords “Giant BCC, metastatic BCC, and aggressive BCC.” The latest literature review addressing these topics, the study published by Zoccali et al. in 2011 was reviewed and the content
was updated with studies published from 2011 to 2014, as well as those not included in this study.

The case reported in our study with a GBCC localized in the lumbosacral region with no local or distant metastasis was examined for its development during and after the treatment, and the literature was retrospectively screened for localizations in which GBCC are encountered. The case report presented in our study was prepared in line with the guidelines of the Declaration of Helsinki and patient’s consent was obtained for using the information and photography of the procedure.

**CASE PRESENTATION**

Our study reports the case of a 52-year old male patient with a giant exophytic ulcerated lesion that started in the lumbosacral region and grew in a course of 12 months (Figure 1). No predisposing factors with respect to a basal cell carcinoma (regional exposure to sun, arsenic, chemical carcinogenesis, radiation, thermal or chemical burn, chronic trauma) were identified in the patient’s evaluation. The patient had no familial history of skin cancer.

The patient’s examination revealed an ulcerated erythematous lesion of 16 x 12.5 cm (200 cm²) extending from the third lumbar spine to the perianal region. Radiological assessment (CT) did not report any bone invasion (Figure 2), and no additional pathologies were identified in the overall physical examination. Dermatoscopic examination found the lesion to be consistent with a basal cell carcinoma with no distant metastasis, but the tumor had considerably grown as an outcome of the patient’s fear of surgery. An excision procedure with frozen section was scheduled. After the margins of the lesion and the type of the pathology were confirmed using the frozen technique, the defect was reconstructed with partial-thickness skin grafting.

Pathology examination of the 18 x 14.5 cm specimen containing cutaneous and subcutaneous adipose tissue reported basal cell carcinoma of both ulcerative and infiltrative types. The tumor, as consistent with aggressive BCCs, presented irregular contours, invasive tumor islands in the dermis, and fibrosis and mitotic cells in surrounding tissues (Figures 3a, b).

The patient was discharged on day 5 following the surgery. No problems were encountered in the clinical follow-up examinations (at postoperative months 1, 3, 6, and 12) and no signs of recurrence or metastasis were identified in imaging (Figure 4).

**DISCUSSION**

Basal cell carcinoma is the most frequently encountered type of carcinoma in skin cancer with a prevalence of 65-80% in non-melanoma type skin cancers. BCC was first described by Jacob in 1827 as rodent ulcer, and later differentiated from epithelial tumors by Krompecher in 1903. BCCs arise from pluripotent epithelium cells and hair follicles. They often occur in fair-skinned male adults as a result of sun exposure. Their incidence rate is higher among older populations. 95% of the cases are seen to occur between the ages of 40 and 70. While about 93% of the lesions are seen in the head and neck region, they are seen to localize on the nose at a rate of 26%. Prevalence of BCCs localized in the extremities and the torso is reported to be higher in Australia.

The term Giant BCC is used for defining very rare cases of lesions that occur larger than 5 cm in diameter. Invasion of extradermal structures such as muscle, cartilage, or bone are often identified at diagnosis. GBCCs constitute at least 1% of...
GBCCs, too, typically arise from the basal layer and adnexal cells of the epidermis as a result of sun exposure.7 Extensive BCCs can often develop into GBCCs with the contribution of factors such as age (>50), low education level, presence of accompanying health issues (chronic alcoholism, iron deficiency anaemia, depression, Alzheimer’s disease, amyloidosis, or HPV infection), patient negligence, or negligence on the doctor’s side as a result of lack of knowledge.2 Aggressive tumor histology (morpheiform, micronodular, infiltrative) can also lead to the rapid growth of the tumor.2

The exact etiology of BCCs is not known, however, exposure to UV radiation is among its frequent causes. Other etiological factors include a specific phenotype (red or blond hair, freckles, Fitzpatrick type 1 or 2 skin), some genetic syndromes (Nevoid BCC syndrome, Xeroderma pigmentosum, Bazex-Dupré-Christol syndrome, Rombo syndrome, multiple hereditary infundibulocystic BCC, familial trichoepithelioma)17, polycyclic aromatic amines, arsenic, x-ray, scar tissue, immunosuppression, sebaceous naevus, family history, and amyloidosis.10 Sun exposure and other predisposing factors were not present in our patient. In many studies smoking has been demonstrated to increase the probability of skin cancer. This was also the case in our patient who had been a heavy smoker for 35 years. Genetic examinations have shown GBCC to be possibly linked to a mutation involving the PTCH gene at the 9q22.3 chromosomal locus, and that this gene plays a role in tumor suppression through an inhibition mechanism of the hedgehog pathway during embryogenesis.18

In our study, (pre- and post-2011) publications not included in the giant basal cell carcinoma literature review conducted by Zoccali et al.16 in 2011 were added and updated. Our search conducted in PubMed on GBCC returned 253 case reports (Figure 5).16,19-42 While in existing literature GBCC is most commonly reported to localize to the face and the back, the head and neck came forth as the top region after the addition of post-2011 literature. This is thought to be a contribution of Arslan et al’s32 2012 study including 34 cases with GBCC in the head and neck region. In the literature review, only five of the cases were identified to be reported in the lumbosacral region.16,35,39 GBCCs were identified to be very rarely-at a rate of 1%-localized to the neck.

Basal cell carcinoma is characterized by slow-growing, non-aggressive lesions. Malignancy potential, recurrence rate and surgical resection margins vary depending on the histological type of the lesion.41 The treatment of BCC includes three categories, namely, surgical, destruction, and medical. Surgical treatment includes primary surgical excision or Mohs micrographic surgery techniques. Destruction techniques include electrodessication, curettage, cryotherapy, and laser phototherapy which can be used in lesions smaller than 2 cm. Medical treatment techniques include radiotherapy, intraloesional interferon injection, and topical chemotherapy techniques. Radiotherapy is the most effective medical treatment technique despite its many side effects. Radiotherapy is occasionally used as a palliative option in BCC. Many studies
have demonstrated that T3 tumors could be cured by 75% when treated with a palliative regimen of 8 Gy given in 3 fractions on days 0, 7, and 21, or with 44 Gy hypofractionated radiotherapy. Radiotherapy is contraindicated in patients with xeroderma pigmentosum, and Gorlin syndrome. In cases that cannot be operated on for various reasons, chemotherapeutic agents such as methotrexate, cyclophosphamide, and 5-fluorouracil (5-FU) are used palliatively. Other alternative treatment forms are imiquimod, photodynamic therapy, and Hedgehog signaling inhibitor (GDC-0449).

The accepted treatment method of GBCC is surgery. The risk of tumor residue in cases of GBCCs—despite being excised with wider margins (10 cm)—is 68%. If the excision and reconstruction are planned as a single-stage operation, the surgical procedure should be performed together with the Mohs or frozen section technique, otherwise a two-stage operation should be preferred in which first the tumor is excised and then reconstruction is performed after the pathology margins are confirmed. Destruction and medical treatment methods can be used in patients who cannot tolerate or who refuse surgery.

Metastasis and death from this disease are rarely seen in patients with BCC. Raszewskiet al. advocated that immunodeficiency and stromal damage were required for metastasis. Rare sites of metastasis from BCC are the lymph nodes, the lungs, the liver and, the spleen. In cases suspected of metastasis, PET CT and scintigraphy should be among the methods considered for imaging. Only a few type of BCCs are invasively aggressive and have metastatic capacity. While metastasis rates are 2% in lesions larger than 3 cm, 25% in lesions larger than 5 cm, and 50% in lesions larger than 10 cm, in lesions larger than 25 cm metastasis is usually already present at diagnosis. Median survival is about 8 to 10 months in BCCs that are already metastatic at the time of diagnosis.

Basal cell carcinomas usually progress by dermal proliferation, invading extradermal tissues such as muscle, cartilage, and bone. In our patient the dermis and the subdermal tissue were infiltrated by the tumor, but there was no invasion of the muscle or the bone. Given the localization of the lesion and the age of the patient, dermoid cyst, epidermoid cyst, pelvic rectal abscess, chronic granuloma, perianal fistula, metastatic disease, desmoid tumor, soft tissue sarcoma occurrences were assessed for differential diagnosis. GBCC patients’ negligence of an existing lesion, hence failure to apply to a clinic is the major factor leading to massive growth. This tendency is oftenfound to be associated with the patient’s educational background.

CONCLUSION

Giant basal cell carcinomas are very rare forms of skin cancer that often occur as a result of the negligence on the patient’s part. GBCCs constitute less than 1% of all diagnosed BCC lesions, however, it should be born in mind that the probability of metastatic development increases significantly as the lesion grows in size, and that diligence is of vital importance in both diagnosis and treatment considerations.

REFERENCES


34. Manstein CH, Manstein ME, Beidas OS. Giant basal cell carcinoma: 11-year follow-up and seven new cases. Plast Reconstr Surg 2011; 128(5): 1105-6. [CrossRef]


49. Randle HW, Roening RK, Brodland DG. Giant basal cell carcinoma (T3). Who is at risk? Cancer 1993; 72(5): 1624-30. [CrossRef]