Two Giant Basal Cell Carcinomas Presenting Simultaneously in the Same Patient, One Resulting in Lower Extremity Limb Loss

Mark Mainella, DO
W. Tomasz Majewski, MD
Predrag Latkovich, MD
Basil M. Michaels, MD

Giant basal cell carcinomas are a rare form of the most common malignant neoplasm of the skin. They are commonly found on the trunk and display a more aggressive behavior, resulting in local invasion and metastasis. Giant basal cell carcinomas that reach a critical size of 10 cm in diameter almost always present with metastases. We present an 82-year-old woman who presented with two giant basal cell carcinomas of critical size—one that occurred on the lower extremity and resulted in limb loss without any sign of metastasis.

Mainella M, Majewski WT, Latkovich P, Michaels BM. Two giant basal cell carcinomas presenting simultaneously in the same patient, one resulting in lower extremity limb loss. Ann Plast Surg 1998;41: 444–447

From the Departments of Surgery and Pathology, Berkshire Medical Center, Pittsfield; and the University of Massachusetts Medical Center, Worcester, MA.

Received Nov 6, 1997, and in revised form Apr 16, 1998. Accepted for publication Apr 16, 1998.

Address correspondence to Dr Michaels, Department of Surgery, Berkshire Medical Center, 725 North Street, Pittsfield, MA 01201.

Basal cell carcinomas (BCCs) are the most common malignant neoplasm of the skin, occurring in more than 500,000 patients per year in the United States alone.[1] The incidence of BCC appears to be increasing at a rate of approximately 3% per year.[2] Usually patients present with multiple small lesions occurring on sun-exposed areas that are easy to treat and are rarely invasive or metastatic. A small subset of BCCs (<1%) are defined as giant basal cell carcinomas (GBCCs). They commonly present on the trunk and usually behave in a more aggressive manner, often leading to local invasion, metastasis, and death.[1,2] We present a woman with two separate GBCCs—one of which occurred on the lower extremity and eventually resulted in limb loss.

Patient Report

An 82-year-old fair-skinned woman with a past medical history of noninsulin-dependent diabetes mellitus presented to the surgical service with multiple, foul-smelling skin ulcers and complaints of a 13.6 kg weight loss during the past 6 months. The patient stated that she had not seen a physician for more than 40 years. A year prior to presentation she had noticed several small skin lesions on her head, trunk, and extremities that had enlarged progressively. In addition, she noted that the lesions bled occasionally and emitted a foul-smelling discharge. She had hidden the lesions successfully from her family until recently, when they found her lesions because of their odor and persuaded her to be evaluated.

On initial examination, a total of six lesions were observed. The largest and most disturbing were a 17 × 17-cm ulcer on her left lower leg and a 10 × 25-cm ulcer on her left upper back (Fig 1). The left leg ulcer was almost circumferential on the lower third of the leg and had a central necrotic area that had invaded the tibia with surrounding cellulitis. It did not appear invasive to the underlying muscle. The remaining lesions consisted of a 5 × 5-cm fungating lesion of the left chest, a 1 × 1-cm lesion of the right forehead, a 6 × 4-cm ulcerated lesion of the right leg, and a 3 × 2-cm lesion of the right neck. There was no lymphadenopathy present on physical examination.

Preoperative evaluation, including chest radiography, abdominal computed tomography, and colonoscopy, was negative for any metastatic disease or second primary malignancy. Radiography of the left leg revealed osteomyelitis at the site of the ulcer. Initial laboratory

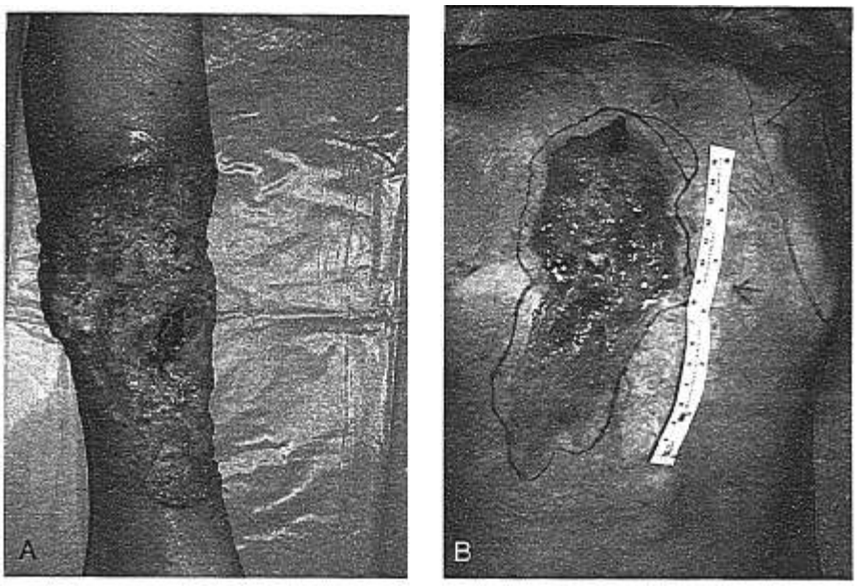

Fig 1. (A) Nearly circumferential ulcerated, lower extremity giant basal cell carcinoma with exposed tibia. (B) Massive left scapular lesion with overlying skin ulceration.

data were remarkable only for microcytic anemia and hypoalbuminemia.

The patient was placed on intravenous antibiotics and taken to the operating room to undergo multiple incisional biopsies. On frozen section, all lesions excluding the left chest lesion (which was read as a squamous cell carcinoma), were read as BCCs. Biopsy of the left cheek lesion revealed actinic keratosis. All lesions were excised with clear margins except for the left lower extremity BCC, which was too extensive and invasive for limb-sparing excision. The multiple lesions were closed by various means: the huge back lesion with a musculocutaneous latissimus dorsi flap, the right forehead lesion with a V-Y advancement flap, and the right leg lesion with a full-thickness skin graft from the right groin. The remaining lesions were closed primarily. The choice of closure was made on the basis of durability. The latissimus dorsi flap was chosen over a split-thickness skin graft because it was felt to be resistant to destruction from direct pressure and abrasion during the healing phase. The right leg lesion was closed with a full-thickness skin graft for the same reason.

Because the patient had been opposed initially to amputation, despite discussion of the consequences, it was decided to treat her with postoperative radiation. Treatment lasted 2 months and consisted of 20 treatments using opposed fields on a Cobalt 60 unit for a total dose of 4,000 cGy. After these initial treatments she received 9 MeV electrons with a 1-cm bolus carried out for a total of 6,000 cGy. Despite considerable reduction in the size of the lesion after radiotherapy, the patient returned with persistent osteomyelitis and a nonhealing ulcer. On that admission she consented to undergo aggressive management of the lesion, and a left below-the-knee amputation was performed. She tolerated the procedure well and was transferred to the rehabilitation unit. In the interim she has required excision of another BCC on the posterior neck. She has been disease free since this time and she walks with a prosthesis.

Pathology

Microscopic examination of the two giant lesions revealed multiple nodules with a lobulated appearance consisting of small keratinocytes and an outer layer of pallisading larger cells (Fig 2A). The stroma was fibromyxoitic with scattered inflammatory cells and characteristic retraction from tumor clusters. Areas of centrolobular necrosis and ulceration of the overlying epidermis were observed. Infiltration of tumor islands in the bone from the extremity lesion was identified in the lesion after amputation (Fig 2B). This histological pattern corresponds with a nodulocystic variant of BCC–the most common form of this tumor (approximately 70% of patients).[3] There was no evidence of squamous or basosquamous cell carcinoma.

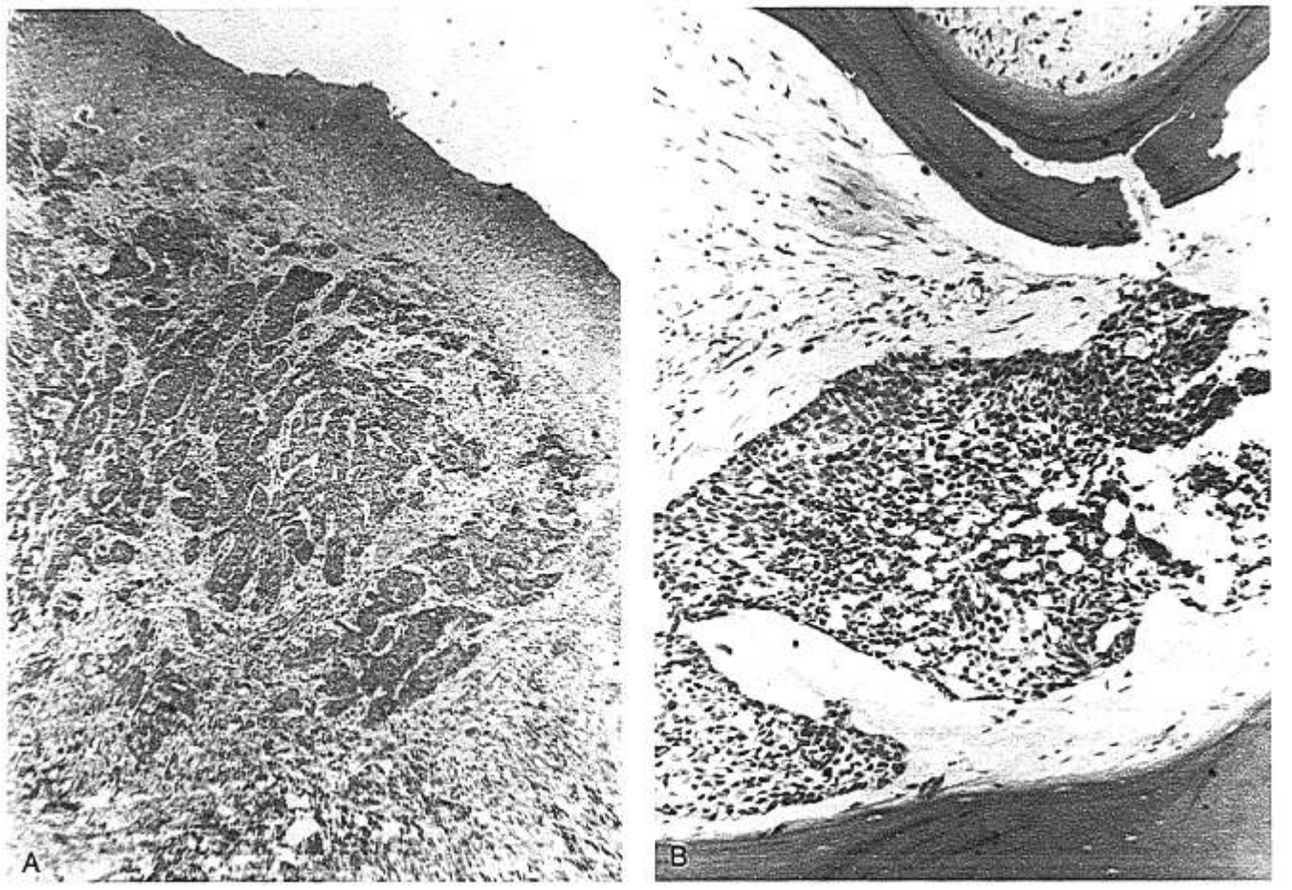

Fig 2. Nodulocystic variant of a basal cell carcinoma (H&E, original magnification × 20 before 93% reduction). (B) Infiltration of tumor islands in the bone from the lower extremity giant basal cell carcinoma after amputation (H&E, original magnification × 40 before 93% reduction).

Discussion

BCCs represent the most common cutaneous malignancy in whites. The reported incidence (500,000–600,000 per year)[4] most likely underestimates the true prevalence of cutaneous carcinomas because most patients are diagnosed and treated in an outpatient setting and therefore are not recorded in a tumor registry. Most of these tumors are small (<1 cm in diameter), well defined, slow growing, and nonaggressive. They occur predominantly on sun-exposed or sun-damaged areas (approximately 80–85% on the head and neck) of fair skinned persons, 95% of them occur in patients older than 40 years, and they present more commonly in men (male-to-female ratio, approximately 2:1).[1,4,5] They arise from the basal cell layers of the epidermis and adnexal structures. There is no known precursor lesion for BCC. Factors considered to be contributing to development of BCC include sun (ultraviolet) exposure, exposure to arsenic, irradiation, coal tar derivatives, and thermal burns.[2] Some researchers indicate importance of genetic factors. Metastatic BCC is very rare (0.0028–0.55%).[6,7]

GBCC is a rare variant of BCC. GBCCs are usually T3 lesions (tumors larger than 5 cm or with deep infiltration of the dermis) or T4 lesions (tumors involving other extradermal structures such as bone, muscle, and cartilage). They are located preferentially on the trunk, although only 10% of all types of BCCs are found on the trunk.[2] They are usually large, destructive, highly malignant tumors with a significantly increased potential for metastatic spread and even death. Patients with GBCCs are more likely to have a tumor that has an "aggressive" histological type (morpheaform, micronodular, or metatypical). However, there is no evidence that GBCCs grow more rapidly than other BCCs. The size of the tumor seems to

be a function of duration rather than rapid rate of growth.[1]

There are several aspects of our patient that make her case unique. First, ours is the first reported case of a patient presenting initially with two GBCCs. Using the definition that has been accepted recently by Sahl and coworkers[2]—a lesion 10 cm or larger in diameter prior to surgery—our patient had two moderate-size GBCCs.

In addition, the location of the massive lower extremity lesion is rare. As stated previously, the vast majority of GBCCs are located on the trunk, and only few have been reported occurring on the extremities. Cruse and colleagues[8] reported a GBCC on the upper extremity, but none have been reported on the lower extremities—yet alone the leg.

Moreover, similar to the patient of Cruse and colleagues,[8] in whom a GBCC resulted in limb loss and metastases, our patient is the only reported patient in whom the limb loss was in the lower extremity without any metastasis. Also, our patient was an exception to the findings of Sahl and coworkers,[2] who found that tumors with areas larger than 100 cm^2 resulted universally in metastasis and often death. In our patient both tumors exceeded this size, and the patient has been disease free with no evidence of metastasis since presentation.

Other findings that were present in our patient and are consistent with other series include iron deficiency anemia and hypoalbuminemia secondary to the intermittent bleeding and weeping of these giant lesions.[2,9,10]

We postulated causes for the size and invasiveness of these lesions. The histological pattern did not correspond with the more aggressive subtypes reported by Jacobs and associates.[11] Nor did our patient have prior exposure to radiation or ingestion of arsenic. Other factors contributing to development of BCCs, such as exposure to coal tar derivatives or thermal burns, were also absent in our patient. The most likely factor contributing to the size of these lesions, as in other reported patients, was neglect.[2,9,10]

There are a few lessons to be learned from our patient. As stated in other reports,[2,12] BCCs may demonstrate very malignant behavior if the tumor size becomes large (>100 cm^2), in terms of either increased metastatic potential and/or local invasiveness. Very large tumors may require extensive surgical treatment, necessitating limb amputation or resulting in major disfigurement. Radiotherapy at best will provide marginal reduction in the size of the lesion, but cytoreduction alone is probably not sufficient to cure a patient. The most common cause relevant to development of GBCC is neglect; therefore, early diagnosis and treatment of BCCs, and close follow-up are strongly recommended. Surgery, either by means of traditional excision or Mohs micrographic surgery, remains a mainstay in the treatment of BCCs.

References

1 Randle HW, Roenig RK, Broadland DG. Giant basal cell carcinoma (T3)—who is at risk? Cancer 1993;72(5):1624–1630

2 Sahl Jr WJ, Snow SN, Levine NS. Giant basal cell carcinoma. J Am Acad Dermatol 1994;30(5 Part 2):856–859

3 Lang Jr PG, Maize JC. Histologic evolution of recurrent basal cell carcinoma and treatment implications. J Am Acad Dermatol 1986;14:186–196

4 Berger DH, Feig BW, Fuhrman GM. Nonmelanoma skin cancer. In: The M.D. Anderson Surgical Oncology Handbook. Boston: Little, Brown and Company 1994

5 Siegle RJ, Wood T. Nonrecurrent primary basal cell carcinoma of the lower extremity with late metastasis. J Dermatol Surg Oncol 1994;20:490–493

6 Snow SN, Sahl WJ, Lo JS, et al. Metastatic basal cell carcinoma—report of five cases. Cancer 1994;73(2):328–335

7 Smith JM, Irons GB. Metastatic basal cell carcinoma: review of the literature and report of three cases. Ann Plast Surg 1983;11(6):551–553

8 Cruse CW, O'Neill W, Ryhack J. Metastatic basal cell carcinoma of the upper extremity. J Hand Surg [Am] 1992;17(6):1093–1094

9 Yamamoto S, Johno M, Kayashima K, et al. Giant basal cell carcinoma associated with systemic amyloidosis. J Dermatol 1996;23:329–334

10 Curry MC, Montgomery H, Winkelmann R. Giant basal cell carcinoma. Arch Dermatol 1977;113:316–319

11 Jacobs GH, Rippey JJ, M Altini. Prediction of aggressive behavior in basal cell carcinoma. Cancer 1982;49:533–537

12 Bianchini R, Wolter M. Fatal outcome in a metatypical, giant, "horrifying" basal cell carcinoma. J Dermatol Surg Oncol 1987;13(5):556–557