Giant Basal Cell Carcinoma: Report of Two Cases and Review of Risk Factors

Greg K. Sakamoto MD and Allan K. Izumi MD

Abstract
GBCCs are rare, tend to be multiple, often develop in sun-protected areas, usually occur in Caucasians without a family history of skin cancer, and have no clear association with immunological or psychiatric factors. Fear may be the underlying factor leading to neglect and chronicity of the tumors.

Introduction
Basal cell carcinoma (BCC) is the most common cutaneous malignancy in the United States, accounting for 65-80% of all reported cases of non-melanoma skin cancers and involves 500,000 cases a year. The lesions occur on sun-exposed areas of the body, with 75% of cases occurring on the head and neck. Factors that affect the development of BCCs include ultraviolet radiation, ethnicity, and genetic predisposition. Less common causes include arsenic, coal tars, x-rays, and thermal burns. Most of the lesions are small, well-defined, and rarely metastasize. BCCs rarely develop into a large tumor referred to as a giant basal cell carcinoma (GBCC), which is variably defined as a tumor larger than 5 cm to 10 cm in diameter.

Case Report
Case 1
A 59-year-old divorced, Caucasian man presented with a large, ulcerated tumor on his left shoulder and smaller tumors on his right arm, left chest, and right thigh. The tumor on his left shoulder developed about 15 years ago, and gradually enlarged, ulcerated, and bled. He was vague about why he never sought medical attention. In December of 2002, while visiting Hawaii, he felt weak and went to a health screening service where he was found to be severely anemic. He was admitted to the Queen’s Medical Center, where he was found to have severe iron-deficiency anemia with a hemoglobin of 7.6 g/dL and hematocrit of 27.9/L. The patient received four units of packed red blood cells. A hematologic work-up to evaluate the iron-deficiency anemia included a colonoscopy, EGD, and small-bowel follow-through, which demonstrated non-specific gastritis and two colon polyps and no obvious signs of bleeding. He had no family history of skin cancer.

Physical examination revealed a pleasant, oriented Caucasian man who appeared psychologically stable with a large 15 X 20 cm fungating, cauliflower tumor with central ulceration on his left shoulder (Fig 1). The tumor had dilated venous extensions with small satellite lesions around the periphery. In addition, he had an 8 X 10 cm fungating, sessile tumor which was friable over his right upper arm, a 5 cm flat, well-demarcated lesion on the right upper thigh, and two smaller well demarcated, friable tumors on his left upper chest measuring 1.5 to 2.5 cm in diameter. A punch biopsy from each lesion confirmed that they were BCCs. The patient was referred to a general surgeon for resection of all tumors. Prior to surgery, an MRI scan revealed that the largest lesion extended down to the level of the acromion and the level of the fascia of the trapezius. The radiologist was uncertain whether the underlying muscle or bones were involved. A CT scan of the chest did not show any intrathoracic lesions. All three tumors were resected with tumor-free margins. The giant BCC of the left shoulder extended into the subcutaneous fatty tissue, but did not involve the underlying trapezius muscle or bone. A reconstructive procedure was done to close the large defect on the left shoulder. The patient received post-operative radiation.

Case 2
A 57-year-old Caucasian man had an ulcerated, bleeding lesion on his right thigh for a year, and smaller lesions on his left and right ear. His medical history was unremarkable, and his family history was negative for skin cancer. On physical exam, the patient was a pleasant, Caucasian gentleman with a normal physical examination except for a large 6 X 6 cm bleeding, ulcerated tumor on his right thigh (Fig 2), and a 2-3 mm tumor on his left anterior apical helix, and a 3-4 mm tumor on his right lower earlobe. The two smaller lesions were curetted and pathology confirmed the diagnosis of BCCs. The larger tumor was biopsied and pathology confirmed an invasive BCC. The large BCC was surgically excised with tumor free margins. The defect was repaired using a split-thickness graft. Prior to surgery, the patient was found to have a mild anemia, with a hemoglobin of 13.7 g/dL and a hematocrit of 40.1/L.

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Discussion

A review of the literature on GBCCs revealed certain common epidemiological factors. Race, multiplicity of tumors, development on sun-covered areas, neglect, and chronicity were common features. Immunodeficiency and genetic predisposition to BCCs in other family members were not consistent factors (Table 1).

Patients with GBCCs often develop multiple lesions, which frequently occur on non-sun-exposed areas. This is in contrast to patients who develop a single small BCC, which usually occur on sun-exposed areas.

Factors which did not seem to play a prominent role in GBCCs included immunodeficiency, genetic predisposition, and anemia. While immunodeficiency influences the development of BCCs and squamous cell carcinomas in HIV and organ transplant patients, there was no trending of immunodeficiency in the patients with GBCCs. Similarly, there was no clear evidence of a familial tendency of developing GBCCs in other family members. Anemia was not a common finding in the case reports of GBCCs, but was reported in several cases. Both of our cases presented with anemia, which was thought to have resulted from chronic blood loss. Histologically, GBCCs tended to be nodular BCCs that were deeply invasive. Biologically, the tumors rarely metastasize, although there were several reported metastatic cases leading to death.

Neglect seems to be the most important factor explaining chronicity, which resulted in the development of these giant tumors. The chronic nature of the tumor most likely results in the development of GBCCs. GBCCs frequently go undiagnosed for at least one year, with some having a duration of 30 years. There is no consistent pattern of educational background, religion, indifference or psychiatric disorders to explain the negligence (Table 1). Based on the lack of information with reference to underlying motivation, it is reasonable to surmise that fear is a basic factor which contributed to neglect. This is supported by the fact that these tumors were primarily in covered areas, which allowed patients to conceal their tumors and their fears. The underlying cause of fear is speculative, and includes fear of diagnosis, treatment, survival, or fear of losing a job or becoming a burden to others.

Surgical excision is the treatment of choice for GBCCs. Most of the reported case studies utilized surgical excision as the initial treatment, followed by a variety of post-surgical treatments such as radiation, skin flaps, and skin grafting.

Our conclusion is that fear may be an important underlying factor in the evolution of GBCCs, which accounts for neglect and chronicity. GBCCs tend to be multiple, develop in sun-protected areas, occur predominantly in Caucasians without a clear familial tendency, and have no clear association with religion, immune status, or psychiatric factors. Surgical excision is the treatment of choice, with a variety post-surgical options.

References


See Table 1, next page
<table>
<thead>
<tr>
<th>Author</th>
<th># of Patients</th>
<th>Race</th>
<th>Duration (Years)</th>
<th>Location of GBCCs</th>
<th>Motivation for Neglect</th>
<th>Multiplicity (Total # of lesions)</th>
<th>Family History</th>
<th>Medical Problems</th>
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<tr>
<td>Manstein et al.</td>
<td>7</td>
<td>NS</td>
<td>6 of 7 pts. greater than 2 years</td>
<td>2 Back, 1 Shoulder, 1 Groin, 1 Lower leg, 1 Neck, 1 Face</td>
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<td>1</td>
<td>Leg &amp; back (GBCCs)</td>
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<td>8</td>
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<td>“Too busy”</td>
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<td>Fe-Deficiency anemia</td>
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<td>Randle et al.</td>
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NS indicates “Not-Stated, NF indicates “No Family History”