Giant Basal Cell Carcinoma

Shayna C. Rivard, MD; Michael L. Crandall, MD; Neil F. Gibbs, MD

ABSTRACT

Servicemembers are often exposed to extreme environments with sun exposure, often laying the foundation for future skin cancer. Basal cell carcinoma (BCC) is the most common of skin cancers. We present the case of a 36-year-old male active duty Seabee who presents with a left shoulder plaque that initially started as an erythematous papule but has now increased to greater than 6cm in the past 10 years and is diagnosed as giant basal cell carcinoma (GBCC). Although only 0.5% to 1% of BCCs develop into GBCCs, there is the potential for metastasis and even death. This article addresses the concerning and potentially fatal diagnosis of GBCC, including your initial impressions and differential diagnoses, available treatment options, and ways to prevent it from ever occurring in our military population.

KEYWORDS: *basal cell carcinoma, giant basal cell carcinoma, enlarging plaque, electrodessication and curettage, UV damage, sun exposure, Seabee, military providers*

Introduction

Servicemembers are often exposed to extreme environments with sun exposure; new recruits are laying the foundation for future skin cancer. We present the case of a Servicemember who had an erythematous papule that increased in size after 10 years and was diagnosed as GBCC. BCC is the most common of skin cancers. Although only 0.5% to 1% of BCCs develop into GBCCs, there is the potential for metastasis and even death. We discuss your initial impressions and differential diagnoses, the specific features you should examine regarding this "rash" that will not go away, and the treatment options that are available. In addition, we describe how you should counsel a patient who has the concerning and potentially fatal diagnosis of GBCC and delineate the follow-up that is needed. This article also addresses ways to prevent it from ever occurring in our military population.

Case Report

A 36-year-old male active duty Seabee presents to your office with a left shoulder plaque that initially started as an erythematous papule but has now increased to greater than 6cm in the past 10 years. He has no significant past medical history. He reports that it occasionally itches and burns but is overall not bothersome. He notes no known inciting event and has no other symptoms. His job occupation includes spending a lot of time in bright sunlight. The patient reports no other history of prior skin disorders.

On examination, he has one large erythematous plaque measuring approximately 6.2 × 4.5cm on his left shoulder with areas of serum crust and scale throughout (Figure 1). On full skin examination, no other concerning lesions are present. The review of systems was within normal limits. Differential diagnoses include BCC, Bowen's disease, eczema, psoriasis, and extramammary Paget's disease.[1,2]

A shave biopsy is performed with pathology results consistent with superficial BCC. As the plaque measures

Figure 1 (Left) *Large erythematous plaque with serum crust and scale on patient's left shoulder.* **(Right)** *GBCC measuring approximately 6.2cm × 4.5cm.*

greater than 5cm, it is classified as a GBCC. Various treatment options are discussed, to include surgery, but due to the size of the plaque, electrodesiccation and curettage with subsequent treatment with imiquimod 5% topical cream (Aldara®) 5 times a week for 6 weeks was chosen. The patient is provided hydroxyzine for symptomatic management of pruritis with plans to follow up in 3 months.

Overview

Basal Cell Carinoma

BCC is the most common of skin cancers and is often described as a scaly plaque with pearly white borders and telangiectasias apparent on close examination or dermoscopy.[1,3,4] BCC occurs in 23% to 28% of white females and 33% to 39% of white males, and these percentages are increasing each year.[2,4,5] Defino et al. (2006) reported on BCC cases between 1994 and 2003 and documented an overall increase of 2.3% per year and, more specifically, of 5.4% per year for those younger than 35 years[6]; additional literature supports a higher increase of 3% to 10% each year.[2,4,5] Due to reported thinning of the ozone layer in conjunction with different recreational and clothing patterns, as well as an increased life span, humans are exposed to greater doses of ultraviolet (UV) rays than in previous centuries.[5] Only 5% of cases are reported to occur in those aged 40 and younger.[4]

Males are also roughly twice as likely to have BCC as are females.[4,7] It appears that males become more easily immunosuppressed with UV doses than do females, suggesting that dose exposure due to occupation or recreation alone may not fully explain higher rates of skin cancer in males.[8] Cytokines are also released and Langerhans cells are altered on UV penetration, both of which may cause immunosuppression.[5] In addition to UV damage affecting DNA via the *p53* gene, *Patched-1* gene, and *sonic hedgehog* pathway in the epidermal basal layer and adenexa, other associations are burns, arsenic exposure, chronic trauma, immunosuppression, radiation, hereditary diseases, and light skin color.[1-3,5,8,9]

Ultraviolet B (UVB) exposure is especially damaging to the dermis, causing direct DNA changes, while UVA causes deeper dermal damage via oxygen species development.[8] Interestingly, evidence supports that chronic exposure to UV rays increases rates of squamous cell carcinoma, while short and intense, albeit infrequent, UV ray exposure resulting in sunburns may lead to increased rates of BCC, as well as melanoma.[8] In addition to UV rays, UV light from welding and sterilization may contribute to overall risk.[8] Despite the fact that 75% of BCCs occurs on the head and neck, the shoulders actually receive roughly two-thirds of UV exposure,[8,9] allowing for the development of BCC as in our case.

Giant Basal Cell Carcinoma

Most cases of BCC are identified early, but, due to neglect, recurrence, or absence of medical care, 0.5% to 1% of BCCs reach measurements of 5cm in diameter or greater and are classified as GBCCs.[3,7,9] In a study of 51 cases of GBCC, Archontaki et al. (2009) reported that the average length of time before diagnosis was 14.5 years with average diameter of 14.77cm, and the age of eventual diagnosis with the highest incidence was in the 60s.[7] GBCCs were most likely to occur on the back, face, and upper extremity at rates of 27.5%, 23.5%, and 13.7%, respectively.[7] Of GBCCs, 0.03% to 0.5% metastasize, typically to lymph nodes, bone, and lung.[3–5,9] Per report, approximately 45% of GBCCs that reach larger than 10cm and 100% of those that reach larger than 25cm result in metastasis.[3,7] This may result in subsequent death.

Mollet et al. (2013) depicts a GBCC found in the late stages after a 62-year-old man sought medical care after 10 years, when a "spider bite" was enlarging. On presentation, the plaque on his back measured 19cm × 19cm and had invaded and exposed vertebrae, with imaging studies concerning for metastases.[3] The patient was placed on palliative care and died 18 months from the time of diagnosis.[3] The reported average time from the diagnosis of metastasis to death ranges from 8 to 14 months.[3,7,9] Archontaki et al. (2009) reported higher rates of 17.6% for metastasis and 17% for mortality in their 51 cases reviewed.[7] Even if death does not ensue, limb amputations may occur due to massive tissue destruction.[4]

Treatment

Basic clinical subtype classification of BCC includes nodular, superficial, fibroepithelial, cystic, morpheaform, infiltrated, and micronodular; the subtype will determine what treatment modalities are effective.[1,2,9] Often, GBCCs are of the nodular, micronodular, infiltrating, fibroepithelial, or morpheaform type, which are considered more aggressive, thus necessitating wide margins of approximately 10mm during surgical excision, often with postoperative radiotherapy.[2,3,7,9] Recurrence rates for BCC of greater than 3cm that are treated with surgery approximate 23%, while in BCC greater than 5cm, a tumor may be inadvertently left behind in 68% of cases.[2,7] Thus, follow-up is paramount; within 3 years, the individual is 10 times more likely to develop another BCC.[1,5,7]

Mohs surgery for GBCC cases has had a success rate of 99%. Although time consuming, this has the additional benefit of being a tissue-sparing technique, which will also consider aesthetic parameters.[7] Conversely and fortunately for the patient in this case, the superficial subtype is considered the least aggressive form of BCC,

although it has still been reported as a subtype for GBCC in the literature.[3,7] Treatment options include, in addition to surgery, electrodesiccation and curettage with imiquimod or fluorouracil application for several weeks afterward.[1,3,5] Clearance from imiquimod therapy is reported as 70% to 94% for superficial BCC of 2cm or less,[2] and recurrence is up to 20% at 2 years.[1] Another treatment option for consideration of superficial GBCC is CO_2 laser, with 80% of cases reported to clear completely after single treatment and recurrence rates of 3.7% to 15.5%.[2] If found, the patient may also benefit from treatment for associated hypoproteinemia and iron deficiency anemia, which have been reported to occur with GBCC.[7]

Differential

The differential diagnoses are important to discuss, as a misdiagnosis by either layperson or provider early on may have led to subsequent disregard by the Servicemember, allowing for the cancer to progress. Common similar diagnoses include Bowen's disease, eczema, psoriasis, and extramammary Paget's disease. Bowen's disease, or squamous cell carcinoma in situ, has is characterized by an erythematous plaque with scale and fissures on sun-exposed portions of the body or mucous membranes, often without the smooth pearly white borders found in superficial BCC. However, clinical distinction can be extremely difficult, thus often necessitating a biopsy.[1] Eczema does have similar scale formation, which BCC can create when serum oozes and forms crusts, as seen in our patient. However, eczema can have patterns of vesicles, erythema, scaling, fissuring, and potentially lichenification, depending on its stage.[1] Psoriasis differs in that the distribution is usually along the gluteal cleft, elbows, knees, and scalp, rather than just one localized area, and often presents with a thicker silvery scale.[1] Finally, extramammary Paget's disease is different in that although rarely (2%) may present on other surfaces, it commonly is found in the genital and perianal areas. Typically, the plaque is erythematous and may weep.[1]

In general, one distinguishing characteristic of superficial BCC is its white pearly ridge and telangiectasias, which may be clearly delineated with dermoscopy or when overlying scale is removed and tension is applied on the adjacent normal skin.[1] When a biopsy of superficial BCC is performed, there are atypical basaloid cells exhibiting a peripheral pallisading pattern with buds of these cells in the epidermal basal layer lengthening into the underlying dermis.[1,3,5]

Prevention and Early Identification

This dramatic case presentation demonstrates the importance of early prevention and identification of cutaneous neoplasms in the military population. Often exposed to extreme environments with sun exposure, Servicemembers are at risk, but they also have the opportunity for annual required health screenings. Due to occupational risks, the military population in general would benefit greatly from increased education. Through increased surveillance, providers could decrease percentages of undiagnosed skin cancers. Incorporating additional survey and review questions regarding sun-protective measures during annual health assessments could help Servicemembers consider their daily habits in occupations that may have increased sun exposure.

BCC is often caused by intense sunburn versus chronic exposure, and typically risk due to past sunburns is established by age 20.[5,8] This means that new recruits, who are most likely employed in the sun, are laying the foundation for future skin cancer. Although increased sunscreen use may decrease the incidence of squamous cell carcinoma, there is evidence to support that this does not cause any decrease in BCCs, thus necessitating the additional need for protective clothing, hats, and shade to minimize potential sunburn.[8] Also, sunscreens are often applied inappropriately and may give a false sense of security.[8] It is important for leadership to be aware of the benefits of rotating Servicemembers through tasks that require ongoing sun exposure as well as ensuring adequate protection against sunburn with personal sunscreen, long-sleeved clothing, and wide-brimmed hats, in addition to the use of shade throughout the day to help decrease the risk for skin cancer.[8] Establishing smoking pits or other areas of congregation that are well covered and shaded, even through the simple provision of awnings for this purpose, would be beneficial.[8] Also, members should be educated on sunglass use in an increasing post Photorefractive Keratectomy (PRK) and Laser-Assisted in situ Keratomileusis (LASIK) population, as roughly 3 million lose vision each year from pathology due to sun exposure.[8]

To truly decrease skin cancer rates in the military population, there is an overall need to change habits through education, such that one chooses to seek shade that leadership provides and encourages instead of basking in the sun without protection.[8] Currently, habits and surveillance methods do not exemplify this, which allowed a BCC to grow to giant proportions in an active duty Servicemember.

Conclusion

In summary, this case raises the concern that despite 95% of BCCs occurring in those older than 40 years, a BCC developed into a GBCC over the course of 10 years in a Seabee who had increased occupational exposure risks. Thus, military providers should have heightened awareness for these neoplasms even in those members in

their mid 20s and seek to educate their military population on the dangers of occupational and recreational sun exposure.

Disclosures

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Disclaimer

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LT Rivard graduated from Vermont College of Medicine in 2011. She completed her transitional year internship at Portsmouth Naval Medical Center in 2012. She briefly served as regimental surgeon for Combat Logistics Regiment 17 at Camp Pendleton, CA, and is now deployed as battalion surgeon for Combat Logistics Battalion 13 on 13th Marine Expeditionary Unit. E-mail: shayna.rivard@lsd49.navy.mil or shaynarivard@hotmail.com.

LCDR Crandall graduated from Saint Louis University School of Medicine in 2007. After an intern year, he spent 4 years as a flight surgeon with the USMC and is currently a second-year dermatology resident at Naval Hospital San Diego in San Diego, CA.

CAPT (Ret) Gibbs is from Naval Hospital San Diego in San Diego, CA. He is program director of the Naval Hospital San Diego Dermatology Residency Program and staff pediatric dermatologist.