

Super giant basal cell carcinoma in an autistic patient: A case report

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Abstract

Basal cell carcinoma is the most common skin cancer in the world and is generally treated when small in size with an excellent prognosis. Rarely, basal cell carcinoma will grow to be larger than 5 cm, at which point they are termed giant basal cell carcinoma. Giant basal cell carcinoma comprises only 0.5% of all basal cell carcinoma, but is associated with impaired quality of life and increased risk of metastasis. When a basal cell carcinoma grows to over 20 cm in size, it is termed super giant basal cell carcinoma. Here, we report a case of both a super-giant basal cell carcinoma and a giant basal cell carcinoma developing over 10–12 years on the upper back and anterior chest wall of an autistic male. Generally, this presentation is associated with neglect on the part of the patient. This case report demonstrates a super-giant basal cell carcinoma developing secondary to patient neglect in the context of comorbid mental illness.

Keywords

Super-giant basal cell carcinoma, autism, cancer, Vismodegib

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Introduction

Basal cell carcinomas are exceedingly common, and are generally identified early with an excellent prognosis. In less than 1% of cases, basal cell carcinomas (BCCs) grow to over 5 cm in size, at which point they are termed giant BCC (GBCC); when they exceed 20 cm, they are termed super-giant BCCs (SGBCC). Generally, these lesions are attributable to patient neglect and loss to follow-up.¹ Due to their size and their tendency for local invasion, these lesions can greatly impair quality of life. There is little consensus regarding the treatment for these SGBCCs, although surgery, chemotherapy, and radiation are the mainstays. Here, we report a case of a patient presenting with an SGBCC which resulted secondary to isolation and neglect.

Case report

A 53-year-old man presented to clinic with two large, friable ulcers on his back and left anterior chest wall. Both had started as small lesions 10–12 years earlier for which he had never sought treatment. The patient was a poor historian who endorsed a history of isolation and avoidance of medical services. His past medical history was significant for severe autism.

On physical examination, the patient was noted to be an alert and oriented Caucasian male. He had an endophytic ulcerating lesion with sharply demarcated rolled borders on his upper back measuring 25 cm (Figure 1). He also had a large

lesion with sharply demarcated rolled borders and necrosis on his anterior chest wall (Figure 2). Both lesions were purulent and friable with serosanguinous and hemorrhagic exudate. The



Figure 1. Sharply demarcated, friable ulcer on the upper mid-back.

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tumor on his back was notable for significant ulceration into the muscular layer, with potential bone involvement. He had no systemic or constitutional symptoms.

A biopsy of the wound edges revealed the diagnosis of BCC (Figure 3).

The patient underwent a PAN-CT scan, which revealed one positive lymph node in the axilla. This ended up being a

reactive lymph node. He was given a diagnosis of stage 3 BCC. The patient was referred to oncology, where he was treated for a stage 3 nonresectable tumor. Treatment plan included tumor debulking with Vismodegib, followed by later assessment about possible further resection. After treatment, the patient showed significant improvement, with shrinking of the tumor to approximately 1/5 of its original size.



Figure 2. Eroding, necrotic tumor on the left anterior chest wall.

Discussion

BCCs are extremely common and generally slow growing with a low risk for metastasis. In rare cases, growth of the tumor to over 5 cm results in a GBCC, an aggressive malignant neoplasm with deep dermal infiltration and involvement of surrounding structures.¹ Literature review found only 11 cases reported.^{2,3} In contrast to BCCs, which are often found in the sun-exposed areas of the head and neck, GBCCs are located predominantly on the back, where they go unnoticed by the patient and can remain hidden under clothing.⁴ There is a greater propensity for metastasis, particularly after reaching a size of 10 cm.⁴

Risk factors for BCC include Caucasian descent, prior history of BCC, and exposure to ultraviolet radiation.

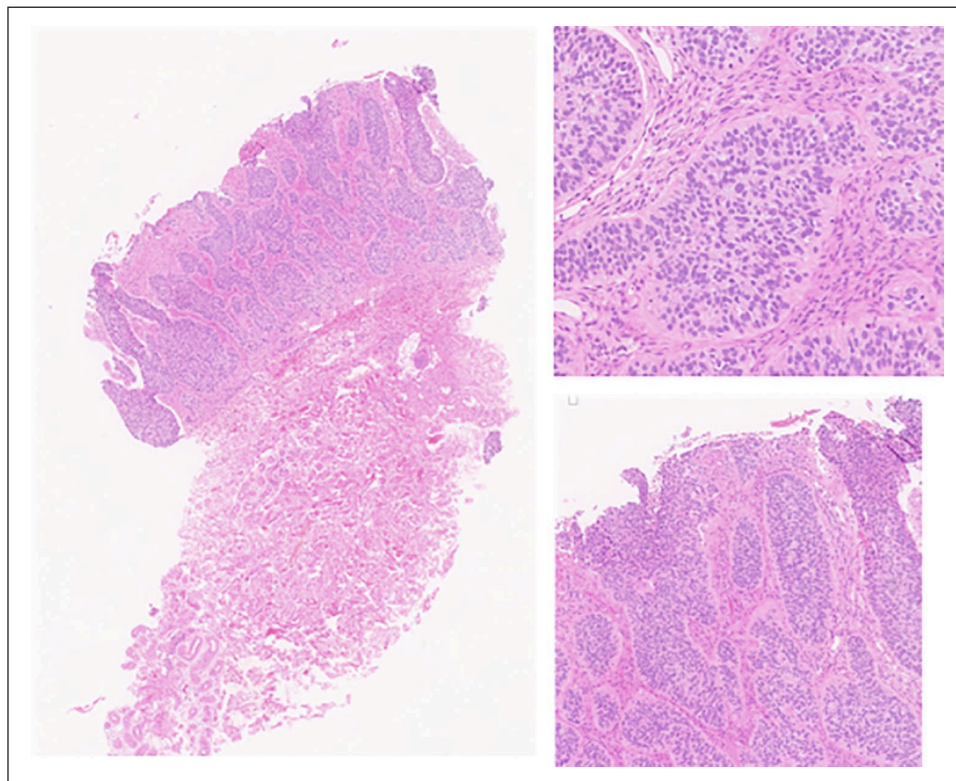


Figure 3. Skin punch biopsy (HE 2×) from the left chest demonstrating variably sized lobules and infiltrating strands composed of basaloid cells with peripheral palisading and focal stromal retraction (HE 10×). Basaloid cells demonstrating variable atypia with large nuclei, prominent nucleoli, and multiple apoptotic bodies and mitotic figures including atypical forms (HE 20×).

These risk factors are shared by GBCC, but larger tumors have additional risk factors including an aggressive histological subtype, recurrence after previous treatment, a history of radiation exposure, or a history of neglect.³ They are most often reported in those with low socioeconomic status, and physical or mental disability which impedes access to health care.⁴ Autism has been found to obstruct use of healthcare through several mechanisms, including fear or anxiety, difficulty communicating with the practitioner, and sensory difficulties with the busy clinics.⁵ In the case of our patient, severe autism led to avoidance of medical care and the extraordinary growth of his BCC.

Although there is no consensus on treatment of SGBCCs, mainstay treatment options include surgical excision, radiation therapy, and chemotherapy.² In 2012, Vismodegib was approved for use by the US Food and Drug Administration for locally advanced and metastatic BCCs that were non-operable, or recurred following surgery.⁶ This hedgehog pathway inhibitor was shown to have response rates of 30% for metastatic BCC and 43% for locally invasive BCC.² Although typically used as a monotherapy or adjunct therapy following surgical resection, Vismodegib has been successfully used for tumor debulking prior to surgery.^{7,8}

Conclusion

This report presents a unique case of non-operable SGBCC. Use of the hedgehog pathway inhibitor Vismodegib was effective in decreasing tumor bulk, allowing for the possibility of future surgical resection.

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M.A.H. managed the patient. E.H. wrote the first draft of the manuscript. M.A.H. made critical revisions.

Declaration of conflicting interests

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

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Informed consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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