

# A rare case of super giant basal cell carcinoma

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## INTRODUCTION

Basal cell carcinoma (BCC) is the most common skin cancer in the world, with 750,000 cases reported annually in the United States alone.<sup>1,2</sup> Because they are typically identified early, it is rare to see BCCs grow beyond 5 cm in diameter; when this does occur, the term *giant basal cell carcinoma* applies. Furthermore, if beyond 20 cm in diameter, the lesion is then termed *super giant basal cell carcinoma*.<sup>3</sup> Because of the rarity of such extensive lesions, there is no consensus on treatment. Although our patient ultimately did not receive therapy for this lesion, similar cases have been treated with surgery and radiation as the mainstays of therapy. However, recent development and use of vismodegib has provided a less-invasive alternative to consider. This case report illustrates a case of a super giant BCC that developed in a modern western society secondary to neglect and poor follow-up.

## CASE PRESENTATION

The patient is a 70-year-old well-educated, accomplished artist and sculptor who presented with intractable diarrhea and malaise. At the time of examination, he had a large bath towel taped to his back with an underlying necrotic lesion with sharply demarcated, rolled borders measuring 20 × 25 cm (Fig 1). The wound, present for more than 35 years, initially started as a “spider bite” on his left upper shoulder in 1978, slowly expanding until he sought medical attention in 1983.

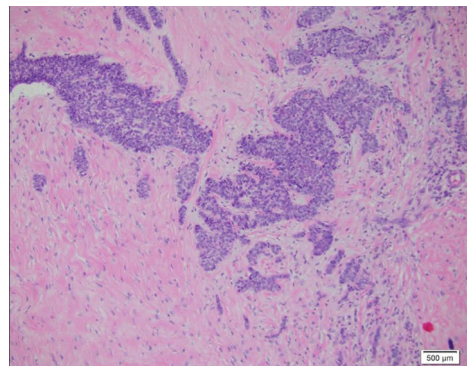
At that time, his primary care doctor diagnosed the lesion as BCC and excision was attempted. The patient claimed the site never healed, and he was lost to follow-up. Over the next decade, the site continued to expand with frequent bleeding and purulence. He did not seek medical attention for the

*Abbreviation used:*

BCC: basal cell carcinoma



**Fig 1.** A 25- × 20-cm BCC with necrosis and oxidation of muscle tissue. Note the friable, rolled borders.



**Fig 2.** Large islands of basaloid cells retracting from surrounding skeletal muscle and stroma; indicating deeply invasive BCC. (Hematoxylin-eosin stain.)

wound during this time because of a “busy schedule” of sculpting and teaching.

In 1995, he moved to a new region of the United States, prompting him to seek medical advice

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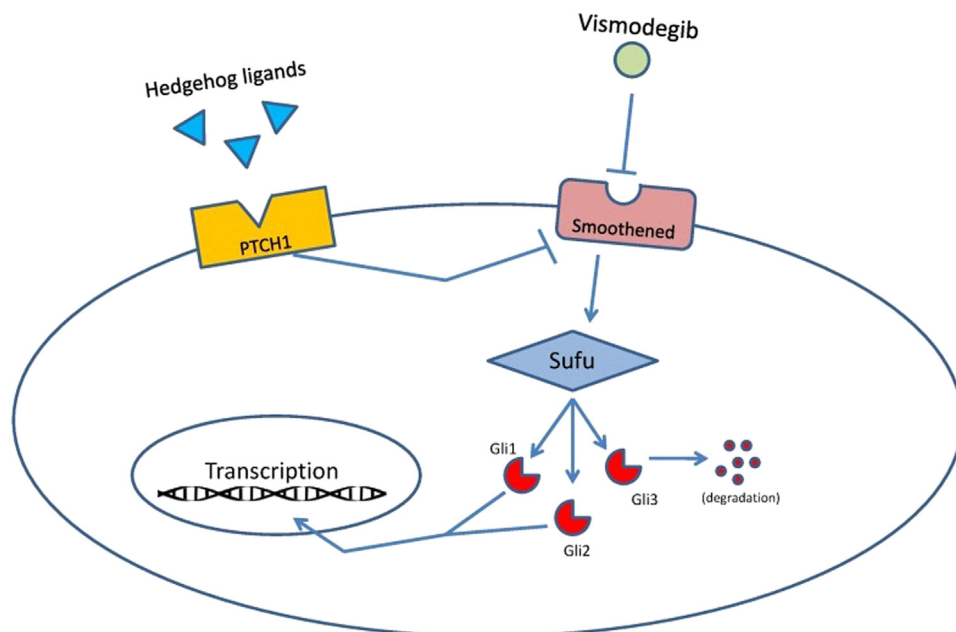
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**Fig 3.** Illustration of the Hedgehog pathway under normal circumstances (no malignancy).

regarding this expanding lesion, now roughly 10 cm in diameter. He sought a more holistic approach, as he believed traditional medicine had failed him, and saw a local chiropractor who began treating the lesion with spinal manipulation and a blue light. After months of poor results, the patient was referred to a physician for evaluation. He was seen by a dermatologist who biopsied the lesion and made the diagnosis of BCC (Fig 2), but the patient declined further interventions, as he felt he was treated poorly by the practice.

Over the next 20 years, the lesion continued to grow, and the only treatment he received was blue light therapy and spinal manipulation from his chiropractor. In July of 2013, he fell ill with headache, diarrhea, and lethargy, and the super giant BCC was rediscovered. At this time, the wound edges were biopsied, showing an infiltrative BCC with skeletal muscle invasion. A computed tomography scan found a mass in the liver consistent with probable metastasis. The patient refused liver biopsy; therefore, metastasis was assumed but never proven. Because the patient was a poor surgical candidate and lesion was too large for complete excision, the oncology department recommended the patient be treated with vismodegib. Unfortunately, the patient died from complications of cancer before the medication was started.

## DISCUSSION

Although typically an indolent, slow-growing cancer, BCC can become aggressive and locally

invasive if left untreated. Giant BCC only accounts for 0.5% of BCCs and super giant BCC is exceedingly rare.<sup>4</sup> Literature review found only 9 previously reported cases.<sup>4,5</sup> These lesions are most commonly found on areas covered by clothing and typically expand because of ongoing neglect by the patient.<sup>6</sup> Archontaki et al<sup>7</sup> published a review of 51 cases of giant BCCs (>5 cm) with the risk of metastases estimated around 6%. The review also documented a significant increase in mortality in patients with metastases.

Previously, treatment options for giant BCC were limited to surgical excision, radiation therapy, and chemotherapy. Vismodegib, a hedgehog pathway inhibitor, acts directly on the G protein–coupled Smoothened receptor. This unique therapy was approved by the US Food and Drug Administration in 2012 for locally advanced and metastatic BCC.<sup>2</sup> With the Smoothened receptor inhibited, neither downstream signaling to the protein Sufu nor release of Gli proteins can occur. This leads to decreased Gli1 and Gli2, which are strong activators of transcription of basal cells. Furthermore, because Sufu is not activated, degradation of the transcription-inhibiting Gli3 protein does not occur and allows Gli3 to function at a baseline inhibitory level (Fig 3).<sup>8</sup> Of note, the beneficial biological properties of vismodegib occur regardless of PTCH1 input. Response rates were measured at 30% and 43% for metastatic and locally advanced BCC, respectively. Median duration of treatment was 7.6 months.<sup>9</sup> Although response rates remain low, one must

consider that this treatment option offers a chance of tumor reduction or clearance for those who might otherwise have no options for treatment.

## CONCLUSION

Giant BCCs greater than 20 cm in diameter are exceedingly rare; we report the tenth case found in the literature. Treatment is often difficult; metastatic rates and mortality dramatically increase with these large lesions. A relatively new therapy, vismodegib, has proven to be an option for some patients in whom treatment may not have previously been available or beneficial for metastatic and locally aggressive BCC.

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