


## CASE REPORT

# Neglected Fungating Giant basal cell carcinoma: A case report and literature review

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## Key Clinical Message

Giant fungating BCC is rare and aggressive. Early health-seeking behavior may result in positive outcomes.

## Abstract

Fungating giant basal cell carcinoma (BCC) is a rare and aggressive form of BCC infrequently reported in the literature. We present a giant BCC case in an old female from a rural area with a poor socioeconomic profile.

## KEYWORDS

basal cell carcinoma, BCC, case report, giant basal cell carcinoma

## 1 | INTRODUCTION

Giant basal cell carcinoma (BCC) is infrequently reported in the literature, with an occurrence rate of 0.5%–1% out of all BCC types, hence representing a rare oncological form.<sup>1</sup> An estimated age-adjusted prevalence for BCC is 343 per 1,00,000 persons per year.<sup>2</sup> There is male predominance among giant BCC with a mean age of diagnoses  $73 \pm 11$  years.<sup>3</sup> Clinically, when the size is more than 5 cm, along with histological BCC, it confirms the diagnosis of giant BCC.<sup>1</sup> BCC is very unlikely among Fitzpatrick skin type 4–6 and is particularly common among Fitzpatrick skin type 1–3, with a 30% risk for lifetime development of BCC.<sup>4</sup>

How giant BCC develops is unclear, but delay in seeking treatment could be the primary cause, as seen in our case. Negligence converts the small BCC into the giant one, a

rare variant that infiltrates the dermis and frequently involves underlying structures, often leading to metastasis and death, but death due to BCC is rare.<sup>5,6</sup> There is no clear guideline for adequately managing a giant BCC, but the surgical approach is the most common. We report a case of a 76-year-old female patient diagnosed with fungating giant BCC arising from negligence.

## 2 | CASE HISTORY AND EXAMINATION

A 76-year-old female presented at the Tribhuvan University Teaching Hospital with a fungating growth in the lateral canthus of her left eye for 8 months. According to the patient, initially, there was an ulcer over her forehead. After a month, concurrent new lesions appeared

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over the nasal bridge and lateral canthus of the left eye. The patient states that the lesions present over the lateral canthus of the left eye progressively increased to the present size over the period of 8 months, with associated purulent discharge and bleeding for the past 6 months. There was no associated fever, pain, difficulty in vision, movement of eyes, or any nasal, and ear symptoms. She had no known comorbidities, did not recall a positive history in her family or have a similar past history. She is a smoker and occasional alcohol drinker. She did not recall going to the hospital for any medical issues and did not report being on any medications or having a history of radiation exposure.

She initially visited the local district hospital and was referred to our center's plastic surgery outpatient department. She was conscious, cooperative, and oriented to time, place, and person. She did not have pallor, clubbing, or lymphadenopathy. Her skin type was Fitzpatrick type 2. Evaluation confirmed the presence of hypertension and hypothyroidism, and other lab findings were within normal limits. Her other vitals were stable. On local examination, there was a baseball-sized fungating, exophytic,

necrotic mass with bleeding areas producing an unpleasant odor. On palpation, the mass was non-tender with woody induration, non-pulsatile, and had no mobility. The mass measured approximately 10×10 cm, extending from the lateral canthus of the left eye to 1 cm in front of the left ear, and the vertical extension was within the temporal region. The overlying skin had irregular bumps with well-defined margins. There were two lesions over the forehead, each measuring 1×1 cm; one had ulceration, and the other had a scab. The ulcerated lesion on the nasal bridge was approximately 2×2 cm, with a well-defined margin and irregular surface extending on either side (Figure 1).

### 3 | METHODS

Clinically, a provisional diagnosis of giant BCC was made. She was referred for ophthalmic, Ear, Nose, and Throat (ENT) evaluation and had no visual or ENT involvement. Before proceeding to surgery, a computed tomography (CT) scan was performed. A whole-body plain CT,



**FIGURE 1** Exophytic growth of giant basal cell carcinoma with ulcerated lesions over forehead and nasal bridge (front and lateral view).



**FIGURE 2** Wide local excision of the lesions (front and lateral view).

especially considering the tumor's giant size, did not reveal any metastasis or neoplastic infiltration of the muscle fascia. There were no associated bony lesions or focal lesions of the brain.

Therefore, a wide local excision with 2 cm margins was performed, and the tumor was removed entirely under general anesthesia (Figures 2 and 3). The excised skin defect was covered with a thick split-thickness skin graft from the right upper thigh (Figure 4). The histopathological finding of the excised mass from the left temple was



FIGURE 3 Resected exophytic growth.

compatible with BCC with squamous metaplasia, while from the nasal bridge and forehead showed squamous cell carcinoma without lymphovascular or perineural invasion and with tumor-free margins (Figure 5). A post-operative ophthalmic examination was similar to the one before the surgery. As the patient was from a rural terrain, hence decided to dress in a nearby district hospital. After that, the patient was lost to follow-up.

#### 4 | CONCLUSION AND RESULTS

Fungating giant BCC is a rare and aggressive form of BCC infrequently reported in the literature. It often results from patient factors and the tumor's nature. There is no clear management guideline, and surgery is the primary treatment. Other treatment modalities may be unadoptable due to cost or availability.

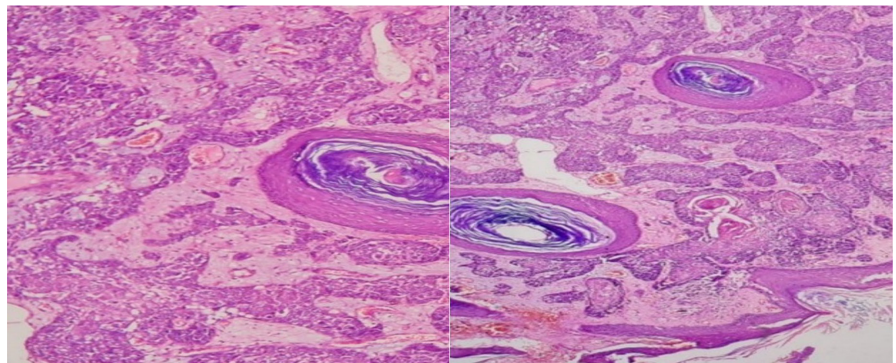
#### 5 | DISCUSSION

BCC is the most common skin malignancy, with a slow growth rate of 1 mm in diameter per year.<sup>7</sup> According to the American Joint Committee on Cancer, a tumor larger

FIGURE 4 2nd post-operative day following split thickness skin graft (front and lateral view).



FIGURE 5 Hematoxylin and eosin (H and E) stain under 100× magnifications shows a basaloid cell nest infiltrating into the dermis with peripheral palisading and retraction artifacts with no perineural or lymphovascular invasion. The stoma shows abundant mucin, and a few keratin-filled cysts are also noted.



than 5 cm in diameter is considered giant cell carcinoma, and only 1% reaches this size.<sup>1</sup> Giant BCC is a rare and aggressive BCC.<sup>1,8</sup>

There is an interplay between environment, patient, and genetic factors for BCC. The modifiable risk factors include exposure to ultraviolet radiation either through occupational exposure or multiple sunburns from an early age, whereas the non-modifiable risk factors include aging, positive family history, genetic factors such as mutations of the patched tumor suppressor genes (PTCH 1 and 2). Other risk factors include Fitzpatrick skin type 1 and 2, a light eye color, blonde/red hair, freckles, photosensitizing medications (tetracycline, hydrochlorothiazide, and statins), carcinogens (arsenic), syndromes (basal cell nevus syndrome, nevoid BCC syndrome, Gorlin–Goltz syndrome, xeroderma pigmentosum, Bazex–Dupre–Christol syndrome), skin pathologies (Kaposi sarcoma, Herpes Zoster, oral candidiasis, hairy leukoplakia), human immunodeficiency virus infection (HIV), organ recipient.<sup>9</sup>

How BCC gets converted to giant BCC is not clear but purported to be associated with several patient factors such as rural residency, alcoholism, professions such as farmers, builders, etc., psychophysical impairments, low socioeconomic status, poor social or family support, poor educational background, all of which contribute to neglect and delay to seek medical advice. Our patient was from a rural residency, belonged to a low socioeconomic group with a low family and social support, and had a poor educational background, contributing to neglect.<sup>10</sup> Other purported causes of developing giant BCC are patients who previously received inappropriate or inadequate treatment, intrinsic aggressive nature, high-risk sites (midface, ear), aggressive histological features (subtype such as morphea form, micronodular, metatypical), radiation exposure, refractory or recurrent even after appropriate treatment.<sup>10</sup> UV exposure in sun-exposed areas is the most common risk factor among the mentioned risk factors. However, BCC can occur in sun-protected areas such as an oral cavity and vulva.<sup>9</sup>

In our patient, the evolution time was 8 months, per the study by Aguilera et al., where they studied 115 patients of giant BCC with  $96 \pm 86$  months (range 6–432 months) of mean evolution time for giant BCC. The head and neck are the most common locations for giant BCC, followed by the trunk, superior, and inferior extremities.<sup>3</sup> In contrast to BCC, giant BCC tends to occur in inaccessible areas covered by clothes, which delays health care visits; in our case, despite being the face area, the patient presented late.<sup>5,11,12</sup> Males have a higher probability of presenting as giant BCC.<sup>3</sup> Patients with BCC may present with enlarged non-healing lesions that may ulcerate or bleed, which are associated with pruritus or giant fungating masses, like in

our case.<sup>1,13</sup> Due to intermittent bleeding and serum loss, anemia, and hypoproteinemia may occur in patients with giant BCC.<sup>14</sup>

To select the most appropriate treatment options and yield better outcomes with a low recurrence rate, the diagnosis and characterization of BCC must be based on the clinical imaging and histopathologic features of tumor mass.<sup>9</sup> Clinical and histopathological features confirm the diagnosis of giant BCC.<sup>1</sup> Dermoscopy is now complemented by novel in vivo diagnostic tools such as optical coherence tomography, Raman spectroscopy, or terahertz pulse imaging to improve diagnostic accuracy and provide tumor depth and lateral margins without invasive techniques.<sup>9</sup>

BCC is one of the most common malignant non-melanotic skin cancers and, as in our case, can coexist with SCC and present as the basosquamous BCC.<sup>9</sup> Considering the histological variant, the most common low-grade histological subtypes are superficial, followed by nodular, adenoid, etc., which has a low recurrence rate, and among the high-grade, the most common is infiltrating, followed by mixed pattern, basosquamous, and morphea form.<sup>3,9</sup> Superficial histological subtypes are more frequent in the BCC groups, while giant BCC has infiltrative histology.<sup>10</sup> Hence, giant BCC is a rare variant that infiltrates the dermis and frequently involves underlying structures, often leading to metastasis and death.<sup>5</sup>

Surgery is the treatment of choice for giant BCC.<sup>15</sup> Small BCCs of size 1–2 cm usually require an excision margin of 3–5 mm,<sup>10,16</sup> while giant BCCs require a wider excision margin of normal tissues (10 mm).<sup>5,9</sup> Either a one-step or two-step surgical procedure is the standard approach. The clinical outcome is inferior in the case of one step approach as the tumor clearance is not visualized; hence, the use of Mohs microscopic surgery has 5 years tumor free rate of 99% for primary and 95% for recurrent BCC when compared with other surgical modalities such as simple excision, electrodesiccation, curettage, cryotherapy, etc.<sup>12</sup> Two-step procedure after the histopathological report is also practiced where the defect is closed via split-thickness skin graft or free flap transfer.<sup>17</sup> For giant BCC, reconstruction is challenging if the lesion is present in aesthetically sensitive areas like ours. The role of the latissimus dorsi musculocutaneous flap, either free or pedicle flap, has been helpful for the reconstruction after extensive surgery with complex defects exposing the nerves, bone, and other vital structures, etc.<sup>12</sup> In our case, we used thick split-thickness skin graft extracted from the upper thigh for the repair of the defect.

Besides surgery, nonsurgical treatments include system chemotherapy with agents such as platinum, cyclophosphamide, methotrexate, bleomycin, vincristine, 5FU, etc., innate immunity stimulators such as immunotherapy, photodynamic therapy, radiotherapy, etc., are

practical for the treatment of BCC.<sup>10,12,18</sup> In the case of inoperable or clinically advanced BCC, sonic hedgehog inhibitor targeted therapy such as vismodegib and sonidegib is the most recent advance in the field of BCC.<sup>10,19</sup> The role of topical imiquimod 5% cream in giant BCC has been shown.<sup>20</sup> Optimal management of giant BCC consists of wide local excision with histologically confirmed tumor-free margins with or without radio chemotherapy and consideration of regional lymph node dissection only in case of lymph node involvement.<sup>1,21</sup>

BCC rarely metastasizes, with a median survival of up to 14 months after the metastasis.<sup>22</sup> It has aggressive behavior, with invasion of deep tissues infiltrating the dermis and involving the underlying structures. It may undergo metastasis, thus carrying a poor prognosis.<sup>1</sup> Tumor size and metastases were correlated, showing 45% of the lesions with sizes more than 10 cm metastasis and 100% of those larger than 25 cm.<sup>23</sup> The giant BCC has a high risk of recurrence, which is supposed to be a 46.9% chance of second primary development within 3.5 years.<sup>23,24</sup> Rigel et al. studied 2960 BCC patients and identified six variables that significantly influenced recurrence rates such as male sex, age less than 50 years, size, number of surgical stages, previous treatment, and anatomical sites such as ear, retro auricular area, and periorbital area with the highest recurrence rate.<sup>25</sup> The recurrence of BCC also depends on tumor features such as its size, location, histologic features, types of treatment, and previous treatment, such as radiotherapy, where the second primary grows larger and becomes more invasive.<sup>26</sup> Our patient was lost to follow-up, which may be due to her socioeconomic status and place of residency; hence, we could not assess the recurrence in this case.

## AUTHOR CONTRIBUTIONS

**Susmin Karki:** Conceptualization; investigation; methodology; validation; writing – original draft; writing – review and editing. **Asmita Parajuli:** Conceptualization; investigation; methodology; validation; writing – original draft; writing – review and editing. **Bhawesh Bhattarai:** Conceptualization; supervision; writing – review and editing. **Khusbu Kumari:** Conceptualization; investigation; methodology; validation; writing – original draft; writing – review and editing. **Kayleigh Anjali Harrylal:** Investigation; validation; writing – review and editing. **Pramish Bhatta:** Writing – review and editing. **K. C. Milan:** Writing – review and editing. **Samit Sharma:** Conceptualization; supervision; validation; writing – review and editing.

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## CONFLICT OF INTEREST STATEMENT

The author(s) declare(s) that there is no conflict of interest regarding the publication of this paper.

## DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author, upon reasonable request.

## ETHICS STATEMENT

Our institution does not require ethical approval to report individual cases.

## CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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