

Received: 2017.06.07
Accepted: 2017.07.18
Published: 2017.12.04

A Rare Case of Giant Basal Cell Carcinoma of the Abdominal Wall: Excision and Immediate Reconstruction with a Pedicled Deep Inferior Epigastric Artery Perforator (DIEP) Flap

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Data Collection B
Statistical Analysis C
Data Interpretation D
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Conflict of interest: None declared

Patient: Female, 82
Final Diagnosis: Giant basal cell carcinoma
Symptoms: Anemia
Medication: —
Clinical Procedure: —
Specialty: Plastic Surgery

Objective: Rare disease
Background: Basal cell carcinoma (BCC) greater than 5 cm in diameter is called giant basal cell carcinoma (GBCC), or super giant basal cell carcinoma if it has a diameter larger than 20 cm. Giant BCC only accounts for 0.5% of BCCs and super giant BCC is exceedingly rare. On account of their rarity, there are no established guidelines for GBCC treatment.

Case Report: We describe a peculiar case of an 82-year-old woman with a GBCC carcinoma of the lower abdominal wall. The tumor was surgically removed with ipsilateral inguinal lymph nodes and the abdominal wall was reconstructed immediately with a pedicled deep inferior epigastric artery perforator (DIEP) flap.

Conclusions: Treatment of giant basal cell carcinoma is often difficult, especially in elderly patients with poor general health and multiple pathologies. The pedicled DIEP flap is rotated to cover the loss of substance without tension, and it is easy to harvest and transfer. This flap allowed a good result without local or systemic complication. We present this report as a reminder of the occasional occurrence of extremely aggressive BCCs. We believe that, especially for rare tumors like these, it is very useful for the entire scientific community to publish these cases and the therapeutic strategies used to treat them.

MeSH Keywords: Basal Cell Carcinoma • Giant Basal Cell Carcinoma • Perforator Flap • Propeller Flap

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/905671>

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Background

Basal cell carcinoma (BCC) is the most common skin cancer. There are about 27 different histological types of BCC [1]. Generally, these BCCs have slow growth and non-aggressive behavior.

Some histological subtypes are more aggressive (such as morpheiform, micronodular, and metatypical) and have been associated with a worse evolution. Metastasis by BCC is rare; the incidence of metastases is estimated to be around 0.1%. As defined by the American Joint Committee on Cancer (AJCC), when BCC reaches 5 cm in diameter, it is called giant basal cell carcinoma (GBCC) or super giant basal cell carcinoma if it has a diameter larger than 20 cm. Giant BCC only accounts for 0.5% of BCCs and super giant BCC is exceedingly rare. Less than 1% of all BCCs reach this size [2].

These tumors typically have slow growth and are almost asymptomatic for many years until they begin to impair quality of life, causing pain, severe anemia, hypoproteinemia due to continuous blood and exudate loss, serious infection, and sepsis.

Giant basalioma, unlike the smaller BCCs, is a rare skin malignancy characterized by aggressive behavior, the tendency to infiltrate deep tissues (e.g., bands, muscles, and bones), and to metastasize. For this reason, these tumors have a poor prognosis. Tumors greater than 5 cm in diameter have a 25% incidence of metastasis [3].

Because of the rarity of these tumors in the literature, there is no large case series of GBCCs. It is therefore difficult to draw conclusions about clinical presentation, histological features, prognostic factors, or treatment strategies from the few clinical cases described.

A literature review suggests that giant BCC typically affects men in their sixties with a history of slow-growing tumor for at least 15 years. Risk factors for the development of giant BCC include delayed presentation, patient neglect, recurrence after previous treatment, aggressive histological subtypes (morpheiform, micronodular, and metatypical), or a history of radiation exposure [4].

Even for giant basal cell carcinoma, the primary cause should be exposure to ultraviolet radiation but the onset in areas not exposed to the sun seems to contradict this statement. It may also occur in patients with basal cell nevus syndromes, xeroderma pigmentosum, albinism, immunodepression, chronic ulcers, or a history of exposure to arsenic, tar derivatives, or x-rays.

On account of their rarity, there are no established guidelines for GBCC treatment. Wide local excision with tumor-free



Figure 1. An 82-year-old woman presented with an enormous, fetid, verrucous, ulcerated abdominal wall mass.

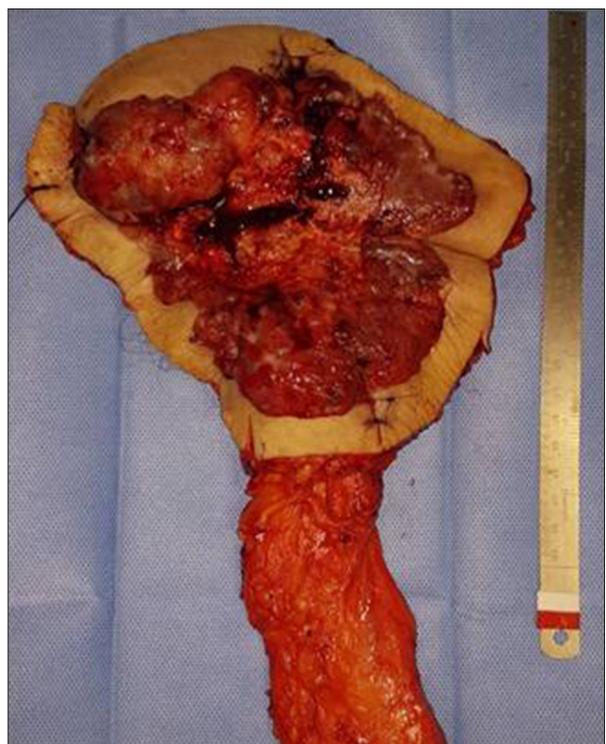


Figure 2. Specimen of the giant BCC, with superficial inguinal lymph nodes.

margins (histologically) is the criterion standard of treatment, but recurrence or metastasis developed in 30–40% of patients, often despite adjuvant radio- and chemotherapy [4]. We describe a peculiar case of an 82-year-old woman with a GBCC carcinoma of the lower abdominal wall. She did not present specific risk factors that favored or induced the onset of this tumor. The tumor was surgically removed and the abdominal wall was reconstructed immediately with a pedicled DIEP flap.

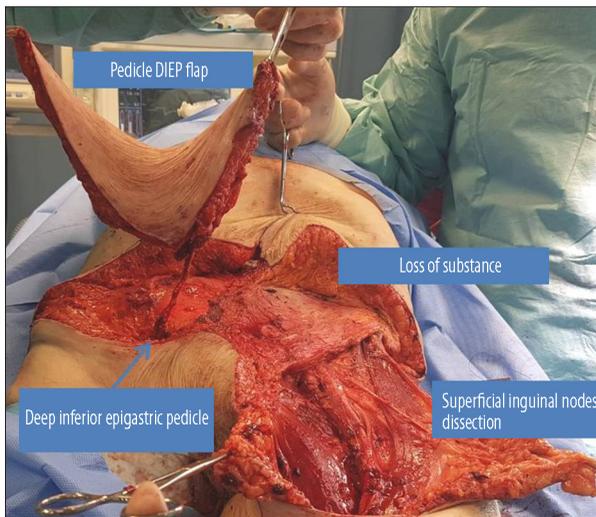


Figure 3. Intraoperative view. The left inguinal node dissection and the loss of substance of the abdominal wall. Pedicled DIEP flap harvested. Highlighting the peduncle of the flap (arrow). The flap is rotated to cover the loss of substance in left hypochondria.



Figure 4. Six months post-op.

Case Report

An 82-year-old woman presented with an enormous abdominal wall mass, 15 cm in diameter (Figure 1), which had appeared on healthy skin about 20 years before and that the patient had never shown to anyone in her family. Following an accidental fall, the patient had reported a femur fracture for which she was admitted to the Orthopedic Department. At the time of the visit, the orthopedist noted at the left hypochondria a verrucous, ulcerated, and fetid mass causing severe metabolic alterations. Blood tests showed neutrophil leukocytosis, anemia (Hb 7 gr/dl), and severe hypoproteinemia. A CT scan demonstrated an infiltration of the abdominal wall, the muscular fascia but not the underlying muscles, without inguinal nodes evolving. No metastases were identified.

Before surgery, blood transfusions were administered until the Hb reached 9 gr/dl. Three days before the surgery, a microbiological test was performed on the tumor, daily boric acid solutions were applied to the mass, and systemic antibiotic therapy (amoxicillin and clavulanic acid, 2.2 gr) was administered. Under general anesthesia, the patient was operated on. Wide and radical local excision with 2.5-cm peripheral margins including the fascia was performed (Figure 2). The superficial inguinal lymphadenectomy was performed in continuity with the tumor. Histologic diagnosis was nodular basal cell carcinoma, with partial adenoid-cystic and scleroderma-like aspects, extensively ulcerated, associated with intense inflammatory infiltrate in the dermis. Ten inguinal lymph nodes were free from neoplastic infiltration. The margins were tumor-free.

The loss of substance was covered using a pedicled DIEP flap, muscle sparing, harvested on the right deep inferior epigastric artery (Figure 3). Using Doppler imaging, a dominant perforator was identified close to the defect. The flap was isolated on a perforator of the medial row of the right deep inferior epigastric artery. Through an incision of the fascia, the perforator was dissected to its origin from the main vessel, which was dissected distally to the right inguinal region. The navel was incised and isolated. The upper abdominal skin areas were detached from the fascial plane for about 10 cm proximally in order to favor the skin closure. The navel was transposed and sutured higher.

The DIEP flap was raised as a pedicled propeller flap, rotated 180 degrees, and placed in the recipient site. The long pedicle of the flap was placed with particular attention to the abdominal muscular fascia, without tension or torsion and without any risk of compression. The muscular fascia was closed and reinforced through a continuous suture and the flap was easily located at the recipient site. The donor site was closed directly. The surgery lasted 3 hours. This considerably reduced the general risks associated with prolonged narcosis. No hernia or bulging occurred after 3 months, and there were no signs of recurrences or metastasis after 6-month follow-up (Figure 4). We present this report as a reminder of the occasional occurrence of extremely aggressive BCCs.

Discussion

Giant BCCs greater than 10 cm in diameter are exceedingly rare. BCC is a slow-growing tumor [1–5]; it grows on average 1 mm in diameter per year [6]. It can then potentially grow to reach large dimensions and infiltrate deep (extradermal) tissues.

Metastasis are extremely rare and reported in less than 0.1% of cases, but the rates of metastasis and mortality increase with increasing tumor size. Tumors greater than 5 cm in diameter

have a 25% incidence of metastasis. Tumors greater than 10 cm in diameter are associated with an approximately 50% increased risk of metastases and death. The prognosis of metastatic BCC is therefore extremely poor [6,7]. The percentage of GBCC metastases reported in the literature is unclear, with wide ranges reported. The rate of metastasis for BCC ranges from 0.003% to 0.55%. According to some authors, the overall incidence of BCC metastases is 0.03% [8,9]. The metastatic rate rises to 1.9% for BCCs greater than 3 cm, to 45% for lesions greater than 10 cm, and 100% for tumors greater than 25 cm.

The most frequent site of metastases are lymph nodes (40–83%) lung, bone, skin, and liver. Metastases occur mainly in males (with a male/female ratio of 2: 1). Average survival with metastasis is 8–14 months [4]. Our literature review found only a few cases [10–15]. BCCs are usually located on the head and neck (80%), and 10% occur on the trunk. These big lesions are most commonly found on areas covered by clothing and typically expand because of ongoing neglect by the patient.

Treatment is often difficult, especially in elderly patients with poor general health and multiple pathologies. As for any epithelioma, surgery is the treatment of choice. As described in the literature, alternative treatments are less effective. Radiotherapy can be useful as adjuvant therapy to control recurrence. Targeted therapy with the hedgehog pathway inhibitor vismodegib [16,17] shows good results in neoadjuvant treatment for advanced or metastatic GBCC, especially in elderly or inoperable patients.

Tumors of the abdominal wall can be treated with a wide local excision but reconstruction can be very complicated. Lesions larger than 5 cm in diameter require clinical free margins of 20–30 mm (or more) because GBCCs have higher incidence of subclinical extension and the risk of tumor recurrence is 68%. Many surgeons prefer to carry out delayed reconstruction, waiting for histological examination to confirm the absence of tumor on excision margins. According to the literature, skin grafts and free flaps (latissimus dorsi, ALT, and TFL free flap) are the main surgical techniques for covering the defect, both in delayed and immediate reconstructions [11–15]. We described a typical case of an elderly woman with a GBCC subjected to radical removal of the lesion and to an immediate reconstruction with a pedicled DIEP flap based on the contralateral deep inferior epigastric artery.

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While its use in free tissue transfer has been widely reported, its use as a local pedicled flap has been more rarely described. In the literature, few reports describe use of the DIEP flap for hip, vulvar, or groin defects reconstruction [18,19] and there are 2 reports of abdominal wall reconstruction [20,21].

The DIEP flap is rotated 180 degrees, as a propeller flap, to cover the loss of substance without tension. The benefits of this technique include, as for all propeller perforator flaps, the avoidance of muscle sacrifice (lower donor site morbidity), the ease of harvest and transfer, and the ability to avoid complications associated with microanastomosis. Another advantage is the plasticity in flap design (“freestyle”). The rapidity of the technique is extremely important, especially in older and comorbid patients, where a prolonged general anesthesia could compromise general health conditions. However, in our patient, the presence of a fracture of the left femoral bone, homolateral to the tumor, led to the exclusion of the use of other flaps (e.g., ALT, TFL, VL) taken from the thigh. Instead, we used the pedicle DIEP flap, which limited the donor-site morbidity to the abdominal region alone, which was the same as the reconstructed site. The pedicled DIEP flap achieved a good result without local or systemic complications.

The postoperative period was regular and uneventful. The patient, during her 6 months of oncological follow-up, did not show signs (either clinical or instrumental) of recurrence or metastatic disease.

Conclusions

We described a case of GBCC in an elderly woman subjected to surgical radical removal of the lesion and to an immediate reconstruction with a pedicled deep inferior epigastric artery perforator (DIEP) flap, based on the contralateral deep inferior epigastric artery. We present this report as a reminder of the occasional occurrence of extremely aggressive BCCs, believing that, especially for rare tumors like these for which there are no treatment guidelines, it is very useful for the entire scientific community to publish these cases and the therapeutic strategies used to treat them.

Conflict of interest

None.

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