CLINICAL VIGNETTE

Giant Basal Cell Carcinoma of the Face with Local Invasion

Jesse Salas, BS[1] and Nasser Samir El-Okdi, MD[2]

[1]University of California Riverside School of Medicine
[2]University of California Los Angeles DGSOM

Introduction

Basal cell carcinoma (BCC) is one of the most common forms of skin malignancy, accounting for approximately 77% of newly diagnosed skin carcinomas.[1] BCC has a relatively low reported incidence of metastasis ranging from 0.0028% to 0.5%.[2] Metastasis, when it occurs, commonly affects vital organs including lungs, bones, liver, and other visceral structures.[3] Patients with BCC originating in the head and neck, accounts for about 85% of BCC occurrences. Their initial metastatic sites are more frequently found in the lymph nodes.[3,4] Early-stage BCC is typically managed through wide margin excision and closure, to prevent invasion of local structures.

When left untreated, BCC can progress to a rare, invasive subtype known as giant basal cell carcinoma (GBCC). GBCC is defined as a tumor larger than 5 cm in diameter.[5] These are more aggressive in biological behavior and have deep tissue invasion and infiltration to extra-dermal tissue, as well as distant metastasis. Previous reviews reported carcinomas with areas greater than 100 cm^2 almost universally result in metastasis or death.[6] Although GBCC represent less than 1% of all tumors, the risk of metastasis is notably increased when compared to smaller lesions that have been treated promptly.[7]

Case

A 51-year old male with history of paranoid schizophrenia requiring prior institutionalization and BCC under his right orbit presented for fatigue and wound evaluation. He was admitted for management of right facial erosion secondary to BCC and profound anemia. The patient had been diagnosed with BCC under his right eye 10 years prior and offered excision of lesion which the patient had declined. Given his history of schizophrenia, he had been placed under his sister's conservatorship care. He was noncompliant with his antipsychotic medications and became voluntarily homelessness after leaving his sister's care. Since the patient's initial diagnosis of BCC, he was recalcitrant with treatment due to his paranoia and fear. Approximately three years prior to presentation the BCC started to erode his face with progression to malodorous drainage and intermittent bleeding. He reported weight loss and fatigue. He had no other significant past medical or surgical history and was not taking any medications at presentation. He had no allergies. Family history included lung cancer in his mother and myocardial infarction in his father. Social history was notable for active tobacco use, occasional alcohol but no current illicit drug use.

He was reunited with his sister and placed under her care and complained of extreme fatigue and difficulty walking and eating for a month. His sister convinced him to come to the emergency room for evaluation. On presentation, vital signs were normal aside for mild tachycardia. His physical exam demonstrated a large cavitation of right cheek with fistula, facial erosion and exposure of a diminutive right ocular globe. There was a noted absence of the right maxilla with pale surrounding skin and a thin non-purulent discharge was oozing from the facial defect (Figure 1). The patient denied any pain from the facial defect.

Admission lab studies included Hemoglobin 3.1, mean corpuscular volume 62.5, platelets 470, reticulocyte count 2.77, iron of 11mcg/dL (percent saturation of 4), ferritin of 7 ng/mL, creatinine 0.30, glucose 181. Wound swab revealed gram-positive cocci in clusters, few yeast, few gram-positive bacilli and moderate WBC. CT with contrast showed right facial soft tissue mass with ulceration and near complete destruction of the entire right orbit, right ethmoid air cells, nasal septum, and right maxilla. There was an extension into the retro-orbital space and the right anterior temporal lobe with mass effect on the underlying brain parenchyma. Diminutive right globe and opacification of right mastoid air cells. CT without contrast of chest, abdomen, pelvis showed no metastatic disease. Follow up MRI without contrast revealed intracranial extension with erosive changes of the right temporal bone and a cystic extra axial lesion along the right anterior temporal convexity. There was mild regional mass effect on the temporal lobe without adjacent edema, tiny focus of FLAIR signals of hyper-densities involving the right superior frontal gyrus and right cerebellum were nonspecific. Underlying parenchymal metastasis was difficult to exclude without contrast (Figure 2).

Wound cultures returned positive for MRSA, pseudomonas, streptococcus agalactiae and lactobacillus. The patient received ampicillin / sulbactam, piperacillin / tazobactam, doxycycline, and ciprofloxacin antibiotic therapy. He received 6 units of packed red blood cells and iron infusions with improvement of

his anemia. Shave biopsy of right upper lip showed nodular type basal cell carcinoma. His case was presented at a multidisciplinary tumor board which included hematology-oncology, radiation oncology, pathology, head/neck surgery and neurosurgery. Plan was made for palliative radiation, and the patient received 10 fractions of palliative radiation therapy while hospitalized. The patient was discharged with referrals for SNF placement with continued wound care and to follow up with hematology and oncology with the plans to pursue vismodegib treatment.

Discussion

Standard treatment for basal cell carcinoma involves excision with 3-10 mm margins depending on size and invasiveness of the carcinoma.[8,9] Therapeutic options for locally advanced BCC were limited in the past due to marginal response to chemotherapy such as methotrexate, fluorouracil, bleomycin, and cisplatin.[10,11] Vismodegib, a smoothened homologue (SMO) receptor inhibitor that blocks the sonic hedgehog pathway in BCC carcinogenesis has been shown to be an effective oral therapy in patients with metastatic BCC, locally advanced BCC, post-surgical, or those who are poor candidates for surgery or radiation.[10] Vismodegib is orally administered once daily until progression of disease halts or patient inability to tolerate adverse effects of medication.[12]

With surgical resection of tumor less than 1.5 cm the 5-year recurrence was reported to be 12% and 23% recurrence with carcinomas above 3cm.[8,13] This patient presented with greater than 5cm BCC with ulceration, oozing, and substantial invasion of local structures. Previous case reports have documented that these the highly treatable BCC may go untreated due to patient neglect, denial of illness, fear of treatment, or lack of access to care.[3,6,14,15] Over time GBCC leads to ulcerations, necrosis of surrounding tissues, invasion of local structures, and concurrent infection with microorganisms, as seen in our case. Median time from onset of BCC to metastasis was 9 years without treatment.[16]

Conclusion

Our patient exhibited GBCC with local invasion without metastasis. Although basal cell carcinoma is highly treatable with excisional surgery, this patient's history of schizophrenia and homelessness were the main barriers in accessing appropriate care. Unfortunately, this is not an isolated case. There have been multiple reports of untreated facial BCC leading to extensive local invasion and loss of facial structures.[3,6,14,15] This patient appeared to be amenable to palliative radiation therapy while hospitalized and initiation of the recommended vismodegib therapy was considered prior to discharge. Barriers included insurance authorization while inpatient and concern for patient adherence to therapy. After discharge, he was lost to follow up and did not pursue further treatments. This case highlights the importance of encouraging patients to undergo early intervention and the importance of follow up with newly diagnosed patients. In patients experiencing homelessness, mental illness or other risks factors for poor follow up, initiation of effective therapy or expedited excision surgery should be considered in addition to addressing mental health issues. Consideration of pursuing LPS conservatorship may be appropriate in patients that lack capacity. Schizophrenia has mortality gap when compared to the general population due to stigma, poor health literacy, under diagnosis, late presentation, and challenges regarding complex decision making. These patients have shorter life expectancies than the general population.[17] Our patient encountered many of these difficulties and deteriorated from an otherwise curable cancer.

Figures

Figure 1: Large right facial erosion due to BCC of face.

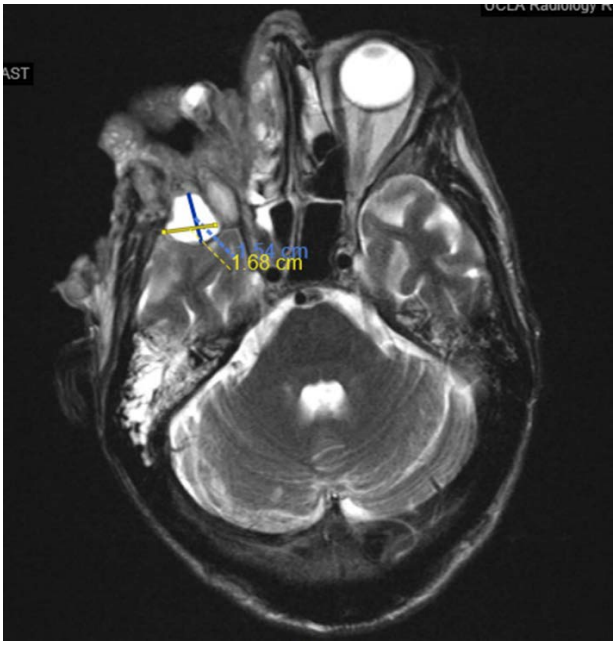

Figure 2: MRI brain showing right facial mass with intracranial extension.

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