Basal cell carcinoma (BCC) is a common skin malignancy comprising 80% of non-melanoma skin cancers (NMSCs). Over 2.8 million cases are estimated to be diagnosed in the United States alone each year. Advanced basal cell carcinomas (aBCCs) are comprised of BCCs that have metastasized to local or distant lymph nodes or organs (mBCCs), or locally advanced BCCs that are extensive and infiltrate vital structures such as eyes, nose or brain (laBCCs). laBCC tumors represent roughly 1.19% of BCCs today. A severe case presentation is presented below.

CASE CLINICAL PHOTOS

INTRODUCTION

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CASE DESCRIPTION

MS is a 69 y/o male with 17 year history of untreated basal cell carcinoma who first presented to our dermatology clinic November 2014. The patient arrived to the office accompanied by his daughter with a towel draped over his face. Upon examination, an extensive ulceration of the entire left face was present, eroding through the left orbit, left maxilla/zygoma and bone of his upper and lower mandible. This was accompanied by complete vision loss of the left eye. The erosion spread through to the right face eroding nasal cartilage/nasal bones and completely obliterating his right globe, orbit and maxilla. (Figures 4a-4c).

According to the records of MS via his previous dermatologist, he first presented to dermatology May of 2012 with a history of biopsy proven basal cell carcinoma that was diagnosed 15 years prior. Patient had denied all previous medical treatments including Mohs, surgery and radiation upon the initial biopsy period. On initial examination in 2012, patient had 25% total vision in left eye and 100% vision in right eye. The ulcer on left cheek measured 4.8 cm x 3.7 cm, L eyelid 3.5 cm x 2 cm and right canthus 2.2 x 2.5 cm with extension through right nasal cavity. (Figures 1a-b)

Patient agreed to take oral Vismodegib upon initial visit in 2012 and was currently on a trial of Vismodegib 1 month upon presenting to our office. While on the medication over the 14 month period the patient lost a total of 75 lbs. Serial CT scans from 2012-2014 showed increasing bone and soft tissue destruction with skull base involvement but no intracranial extension. By June 2013 lesions were increasing in size at a rapid rate of 1 cm per month. MS lost vision in his right eye October 2013. Final measurements in 2014 showed a 13cm x 10 cm left facial ulceration, and 8 cm x 5 cm right medial canthal ulceration. When arriving to our office November 2014 patient stated that he would like to be transferred to hospice with all medications discontinued at this time.

Figure 1a-b: MS at initial presentation May 2012

Figure 2a-b: MS one year after initial presentation September 2013

Figure 3a-b: MS one year after initial presentation April 2014

Figure 4a-c: MS advanced basal cell carcinoma November 2014 (3 years from initial presentation)

DISCUSSION

Basal cell carcinoma (BCC) is a common skin malignancy comprising 80% of non-melanoma skin cancers (NMSCs). Over 2.8 million cases are estimated to be diagnosed in the United States alone each year. This incidence is continuing to rise yearly, as it is now a more common diagnosis than all other malignancies combined.1 BCC is the most common malignancy found in Caucasians, with the fastest growing affected group being women under 40 years of age.2 Most BCC tumors occur on the head and neck and are of almost cosmetic importance.3 Exposure to UV radiation is the greatest risk factor for development of BCC, followed by skin type, family history, prior history of NMSC and immunosuppression.4

Although, fortunately most BCCs are found early and have an excellent prognosis, complex cases do occur as discussed in the previous case. Advanced Basal cell carcinomas (aBCCs) are comprised of BCCs that have metastasized to local or distant lymph nodes or organs (mBCCs), or locally invasive BCCs that are extensive and infiltrate vital structures such as eyes, nose or brain (laBCCs). laBCC tumors represent roughly 1.19% of BCCs, with mBCCs accounting for up to 0.5% of cases.5 Patients presenting with these tumors often present due to a delay in medical attention secondary to severe physical, psychological or financial burden, as seen in the above case. Alternatively, some individuals present earlier with aggressive recurrent tumors refractory to surgical options.

Management of BCC depends on tumor histological type, location, size, comorbidities, previous treatment and preference of patient. Aggressive and infiltrative subtypes are best treated with Mohs surgery.6 Giant tumors defined as those >5 cm in diameter pose a significant challenge to treatment as surgery has high risk of treatment failure and recurrence and can pose significant morbidity and disfigurement.6 In laBCC seen in the above case, at the time of presentation, surgery was simply not a treatment option. In laBCC not suitable for surgery or where surgery is not desired by the patient secondary to impact on quality of life, alternative treatment modalities include radiation therapy; either palliative or curative, chemotherapeutics such as cisplatin (for metastatic disease) or oral Vismodegib.7 No current cytotoxic chemotherapy has been approved for treatment of laBCC.8

Until the past couple of decades treatment options were not based on molecular characteristics that define BCCs. In the 1990s a connection between mutations in the Hedgehog (Hh) pathway and mice was discovered.9 The Hedgehog pathway plays a critical role in cell maintenance for skin cells, stem cells and hair follicles.10 The pathway begins with (Hh) ligand binding to transmembrane receptor PTCH on the cilium of a cell. In the absence of Hh ligand, PTCH is a tumor suppressor and inhibits a protein called Smo. Once binding occurs between Hh and PTCH the Smo signal is activated. This activation leads to a cascade with the end result being proliferation of glioma associated protein (Gli-1, 3). This group of proteins results in oncogenic effect on the cell, aiding cell survival and angiogenesis.10

CONCLUSION

Locally advanced basal cell carcinomas (laBCCs) are extremely challenging to treat. Future therapies may include inhibitors of proteins down-stream to the hedgehog signaling pathway. The case described above shows what a devastating physical, psychological and financial burden these tumors can have. Multidisciplinary care through dermatologists, Mohs surgeons, radiation oncologists, plastic surgeons, psychologists and primary care physicians is needed to provide adequate care for this growing group of patients with challenging BCCs.

REFERENCES