Case Presentation

Recurrent basal cell carcinoma with intracranial invasion: a case report and literature review

Joseph Blackmon MD1, Mac Machan MD1, Anand Rajpara MD1, Robert Beatty MD2

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1University of Kansas Medical Center, Division of Dermatology, Kansas City, KS
2Overland Park Regional Medical Center, Neuroscience Associates of Kansas City, Overland Park, KS

Correspondence:
Joseph Blackmon, MD
3901 Rainbow Blvd. MS 2025
Kansas City, KS 66160
Phone (913) 588-2032
Fax (913) 588-8761
Email jblackmon@kumc.edu

Abstract
Basal cell carcinoma (BCC) is the most common malignancy in humans. We present a man with a recurrent BCC of the scalp that presented as an intracranial tumor 18 years after original excision.

Introduction
BCC tends to follow a benign clinical course characterized by slow local proliferation and very infrequent invasion of muscle, cartilage, or bone. Destruction of the calvarium and invasion of the underlying dura, or brain parenchyma by basal cell carcinoma is extremely rare. We describe a case of recurrent BCC on the scalp with invasion to the level of the dura and compression of the superior sagittal sinus. Additionally, our case is unique in that the clinical appearance of the lesion was rather banal. In almost all prior reports of intracranial BCC, the clinical lesion is exceptionally large and often ulcerated. Furthermore, we briefly review prior cases of intracranial BCC in the literature. This case and review serve as a reminder of the importance of adequate and appropriate treatment of high-risk basal cell carcinoma and underscore the need for physicians and surgeons of all specialties to be familiar with this common cutaneous malignancy.

Keywords: basal cell carcinoma, basosquamous cell carcinoma, calvarium, superior sagittal sinus

Clinical Presentation
A 53-year-old gentleman with a history of a previously excised basal cell carcinoma (BCC) of the scalp vertex in 1994 presented to his primary care physician with an 8-month history of a steadily enlarging lump and associated tenderness (Figure 1) in the region of the prior excision. The original excision 18 years earlier was performed by a plastic surgeon and required skin stretching to complete a linear closure. To fully evaluate the mass, he was referred to a neurosurgeon. On magnetic resonance imaging, a 3.6 x 3.6 x 2.3 cm enhancing soft tissue mass was noted in the left superior parietal calvarium. There was complete destruction of the inner and outer tables of the skull with expansion down to the dura mater. The medial aspect of the mass appeared to involve a small portion of the superior sagittal sinus (Figure 2 & Figure 3). Subsequent magnetic resonance venography showed preserved patency of the sinus.
Neurosurgical intervention was undertaken in an attempt to preserve patency of the superior sagittal sinus and remove as much of the aggressive neoplasm as possible. At operation, care was taken to make an incision to preserve scalp vascularity. The tumor completely eroded the calvarium and extended into the diploic space. A 6 cm craniotomy was enlarged to allow gross visualization of the entire mass. Tumor resection proceeded from the periphery toward the center attachment to the sinus. There was no evidence of frank invasion of the dura and micro dissection allowed complete gross removal from and including the outer dura of the sinus. Bipolar cautery was then used over the superficial surface of the involved outer dura to destroy any remaining microfoci of tumor. Titanium mesh was used to cover the cranial defect. A musculocutaneous free flap was not employed because of anticipated adjuvant radiation therapy. A local rotational skin flap was used for closure.

Figure 1. Scalp vertex demonstrating a red, pink atrophic plaque with surrounding induration.

Figure 2. Magnetic Resonance Image, coronal view: well-circumscribed mass demonstrating intracranial extension through the calvarium to the dura without evidence of parenchymal involvement

Figure 3. Magnetic Resonance Image, para-sagittal view: well-circumscribed mass demonstrating intracranial invasion, extension to the dura and frank compression of the superior sagittal sinus
Final pathological results demonstrated a Stage IV (T4 N0 M0) metatypical BCC, or basosquamous cell carcinoma (Figure 4 & Figure 5), with narrowly clear surgical margins. The patient convalesced well post-operatively and the radiation oncology department was consulted for further medical management of the patient. Given the high rate of local recurrence of T4 BCCs, even with negative surgical margins, adjuvant radiation therapy was recommended and the patient is currently receiving 5000 centi-Gray in 25 fractions with 2 cm margins around the original tumor volume. Radiation therapy was initiated approximately 2 months post-operatively and the patient has tolerated the treatment well.

**Discussion**

BCC is the most common malignancy affecting humans [1], with an incidence in the general population reported at 1% to 2% per year [2,3]. BCCs are generally caused by unprotected exposure to ultraviolet light with additional risk factors being lightly pigmented skin, family history of skin cancer, and immunodeficiency state [4]. More than 1 million cases of non-melanoma skin cancer (NMSC) occur every year in the United States, with greater than 80% of these represented by BCC. Despite greater public awareness of cutaneous malignancies, the incidence of BCC continues to increase by nearly 5% annually [2]. Fortunately, NMSC-related mortality remains very low with an estimated 1000 to 2000 cases per year [2]. These tumors typically present as pink, pearly papules or plaques on the head, neck, or trunk with indurated margins and occasionally central ulceration. Advanced lesions may also present as erythematous plaques or skin-colored nodules [5]. At least 90% of BCCs are cured with the initial therapeutic modality, which includes cryosurgery, photodynamic therapy, topical immunomodulators (i.e. imiquimod), curettage and electrodessication, local excision, and Mohs’ micrographic surgery [2,5]. In most cases, BCCs have an indolent growth pattern. However, years of neglect by the patient may lead to locally invasive disease.

The risk of invasion of vital structures by BCCs is quite low, with an estimated incidence of 0.03% [2,6]. Most of these are located on the head and neck [2]. Previous studies have addressed the relationship between embryonic fusion planes and the depth of invasion, horizontal spread, and recurrence of BCC of the head and neck [6]. The nasofacial sulcus, nasolabial fold, alar groove, and philtrum, all of which are located along fusion planes, tend to have a higher risk of recurrent BCCs versus those at other facial sites [6]. The greater density of nerves and sebaceous glands on the central part of the face may also play a role in the increased risk of recurrence and local invasion of tumor at these sites [6,7]. Computed tomography (CT) is the modality of choice for detecting tumor invasion into bone, which commonly appears as irregular demineralization [6].

There is currently no standard classification scheme for BCCs and at least 26 different subtypes have previously been described [8]. Commonly recognized variants include nodular, superficial, morpheaform, cystic, basosquamous, and micronodular. The nodular subtype represents approximately 60% of all primary BCCs, has a strong predilection for the face, and often enlarges to develop a central ulceration. The superficial variant tends to favor a younger population (average age of 57 years) and is often found on the trunk and extremities. The growth pattern favors a more horizontal direction both clinically and subclinically, which may account for the significant recurrence rate after primary surgical excision [9]. Morpheaform BCC is synonymous with...
sclerosing or infiltrating BCC and typically presents as an atrophic, indurated plaque that is often difficult to differentiate from a scar. Similar to a superficial BCC, the subclinical spread of this variant is difficult to elucidate clinically, often prompting re-excision or primary excision with Mohs’ micrographic surgery [9]. Cystic BCCs have a clear to blue appearance and may produce a clear fluid if punctured. The micronodular variant is a histological diagnosis reserved for those tumors that demonstrate small, deep nesting of tumor aggregates within the deep dermis. This variant is known for its destructive behavior, subclinical spread and high recurrence rate [9]. Finally, the basosquamous, or metatypical variant, demonstrates features of both BCC and squamous cell carcinoma (SCC) both in its clinical and histological appearance. It tends to, therefore, behave more aggressively with a higher probability of metastases (estimated at 9-10%) and local recurrence [10]. All cases of intracranial invasion for which histologic subtype was reported were of morpheaform or basosquamous variants, except for an isolated case of an adenoid cystic tumor.

### Table 1
Previously reported cases of basal cell carcinoma with direct intracranial extension

<table>
<thead>
<tr>
<th>Age (y)</th>
<th>Sex</th>
<th>Location</th>
<th>Size (cm)</th>
<th>Histologic Subtype</th>
<th>Extent of invasion</th>
</tr>
</thead>
<tbody>
<tr>
<td>63</td>
<td>F</td>
<td>Left parieto-occipital scalp</td>
<td>15 X 15</td>
<td>Not reported</td>
<td>Dura</td>
</tr>
<tr>
<td>82</td>
<td>M</td>
<td>Scalp</td>
<td>8 X 10</td>
<td>Morpheaform</td>
<td>Cerebral Cortex</td>
</tr>
<tr>
<td>50</td>
<td>M</td>
<td>Occipital scalp</td>
<td>Unknown</td>
<td>Morpheaform</td>
<td>Cerebellum</td>
</tr>
<tr>
<td>61</td>
<td>M</td>
<td>Frontoparietal scalp and face</td>
<td>17 X 12</td>
<td>Not reported</td>
<td>Cerebral cortex and lateral ventricle</td>
</tr>
<tr>
<td>70</td>
<td>F</td>
<td>Frontal scalp, eyes, nose</td>
<td>20 X 15</td>
<td>Not reported</td>
<td>Cerebral cortex</td>
</tr>
<tr>
<td>47</td>
<td>F</td>
<td>Frontoparietal scalp</td>
<td>Unknown</td>
<td>Adenoid cystic</td>
<td>Cerebral cortex</td>
</tr>
<tr>
<td>36</td>
<td>M</td>
<td>Occipital scalp</td>
<td>1.8 X 1.2</td>
<td>‘Aggressive’</td>
<td>Dura</td>
</tr>
<tr>
<td>57</td>
<td>F</td>
<td>Frontotemporal scalp</td>
<td>Unknown</td>
<td>Morpheaform</td>
<td>Dura</td>
</tr>
<tr>
<td>69</td>
<td>M</td>
<td>Frontoparietal scalp</td>
<td>15 x 15</td>
<td>Not reported</td>
<td>Dura</td>
</tr>
<tr>
<td>57</td>
<td>F</td>
<td>Frontoparietal scalp</td>
<td>11 x 10</td>
<td>Not reported</td>
<td>Dura</td>
</tr>
<tr>
<td>55</td>
<td>F</td>
<td>Frontoparietal scalp</td>
<td>10 x 7</td>
<td>Basosquamous</td>
<td>Dura</td>
</tr>
<tr>
<td>64</td>
<td>F</td>
<td>Frontal scalp, orbit</td>
<td>Not reported</td>
<td>Not reported</td>
<td>Dura</td>
</tr>
<tr>
<td>74</td>
<td>F</td>
<td>Frontotemporal scalp</td>
<td>Not reported</td>
<td>Not reported</td>
<td>Dura</td>
</tr>
<tr>
<td>59</td>
<td>F</td>
<td>Orbit</td>
<td>Not reported</td>
<td>Morpheaform</td>
<td>Dura</td>
</tr>
<tr>
<td>65</td>
<td>M</td>
<td>Frontotemporal scalp</td>
<td>7 x 8</td>
<td>Not reported</td>
<td>Cerebral cortex</td>
</tr>
<tr>
<td>80</td>
<td>F</td>
<td>Frontal scalp, orbit</td>
<td>6.3 x 4.1</td>
<td>Morpheaform</td>
<td>Dura</td>
</tr>
</tbody>
</table>

Our patient developed a recurrent BCC of the scalp with basosquamous histology and intracranial extension at the time of presentation. Notably, our patient’s recurrent lesion was vastly less impressive clinically than many of the previously reported cases demonstrating similar intracranial involvement. It is important to highlight that there has been a different type of aggressive BCC described as occurring on the scalp in young adults under the age of 40 that carries a higher likelihood of local invasion. These rare tumors often advance quickly, invade deeply into local soft tissue and bone, and are resistant to treatment, often leading to death. At the time of surgical excision, the majority of these tumors have invaded through the scalp and into the calvarium;
however, few reach intracranial structures [2]. Table 1 demonstrates only 2 patients that were younger than 50 when diagnosed. Our patient was 53 years old at the time of recurrence, but only 37 years of age at the time of his initial diagnosis.

Treatment of these extensive BCCs can be a difficult task to undertake. Selection of a specific treatment modality depends on the potential for cure, the extent of functional preservation, the desire for a satisfactory cosmetic result [6]. Although the best cure rates are achieved with radical resection, significant deformity may occur depending upon the size of tumor and destruction of the neighboring structures. Other treatment options include radiation and chemotherapy [11]. Recurrences after radiation therapy have been cited as anywhere from 1% to 35% [6]. Systemic chemotherapy may also be considered for an inoperable BCC, but such measures are considered more palliative than curative. Paclitaxel, an antineoplastic alkaloid, has been proven successful in treating aggressive BCCs. A selected case report demonstrated a nearly complete response in a patient with sclerosing BCC after 6 cycles of paclitaxel, cisplatin, and cabecitabine [12]. To our knowledge, vismodegib, the hedge-hog pathway inhibitor approved for metastatic or advanced BCC has not been reported in the setting of intracranial BCC.

In conclusion, our case highlights the rare occurrence of intracranial invasion by a primary cutaneous BCC. Similar to other cases, our patient was over the age of 50. Most patients delayed medical attention and allowed the tumors to grow slowly over several years. However, our case demonstrates that despite only subtle clinical changes to the site, BCC can be a very aggressive and locally destructive neoplasm. Given the overall prevalence of BCCs in the general population, it is of paramount importance to educate patients that not all skin cancer is created equal. Aggressive variants of BCC (morpheaform and basosquamous) exist that warrant more aggressive therapy and subsequent monitoring; these often require multi-specialty care and treatment. The use of vismodegib in management of intracranial BCC should be explored.

References


