Extensive scalp squamous cell carcinoma exposing brain

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Case Report

Abstract

Non-melanoma skin cancer is the most common class of cancer, with Australia having the highest global prevalence. Early detection allows relatively straightforward management, often with surgery or radiotherapy. However, in aggressive subtypes, delayed presentation can lead to extensive local invasion and rarely, this may involve the skull and dura. Multidisciplinary intervention is the most common management of such advanced lesions; there are some clinical scenarios in which surgery is not an appropriate option. Radiotherapy is an alternative definitive therapy that may provide equal rates of locoregional control in less advanced disease, often with excellent palliative cosmetic and functional outcomes. This is especially relevant where surgery is difficult or deforming, a scenario common when lesions involve the head and neck. In addition, due to SCC having high rates of nodal spread and recurrence when incompletely excised, radiotherapy may be a good locoregional adjuvant treatment to surgical excision.

We present a rare case of a very large, long-standing and neglected squamous cell carcinoma of the scalp, which remained localised despite invading through the calvarium into the brain. Following years of poor wound hygiene, the patient presented with dura on show and was eventually found to have a mixed organism wound infection. Surgical intervention was the only definitive treatment attempted and despite extensive resection and intensive care management, the patient died. Given the patient’s complicated co-morbid presentation, we take the opportunity to discuss the balance of treatment goals and the variety of alternative approaches available in difficult cases such as the one presented.

Keywords: Squamous Cell Carcinoma, Central Nervous System Neoplasm, Neoplasm Invasiveness, Surgery, Radiotherapy.

Introduction

Non-melanoma skin cancer (NMSC), comprising predominantly squamous cell carcinoma (SCC) and basal cell carcinoma (BCC), is the most common class of cancer worldwide. Notably, Australia has the highest global prevalence of NMSC, estimated to be 387 per 100000 person-years.¹ The primary site for NMSC is the scalp and head in up to 90% of cases, due to increased exposure to sunlight in a population in which many individuals belong to Caucasian kindreds who have migrated to Australia from the Northern Hemisphere and have lightly pigmented skin.²

The two NMSC subclasses are quite different in their clinical features. Initial presentation is often a nodular lesion in both subtypes, however SCC may be scaly or erythematous and has a tendency to subsequently ulcerate, with raised, irregular edges, whereas neglected BCCs may erode underlying tissues. BCCs are very unlikely to metastasise with an estimated incidence of 0.03% of all presentations.³ In contrast, the local invasion of SCC follows a more aggressive pattern and the risk of distant metastasis is estimated to be up to 3.7%, particularly in poorly-differentiated or recurrent subtypes.⁴ Further, up to 28% of SCC cases involve regional lymph nodes, with a significant impact on mortality.⁵ It is also quite common for less aggressive subtypes of SCC to remain quite localised despite a large size.⁶

While the pathogenesis is multifactorial with a defined genetic component, NMSC is primarily caused by ultraviolet-mediated damage to cutaneous cells. Sun exposure to precursor lesions such as actinic keratosis and Bowen’s disease may play a significant role in the development of these cutaneous neoplasms. With regular medical examination, these precursor lesions should be identified early and excised or ablated. Failing this, NMSCs can usually be identified whilst small and
definitively managed with surgery, radiotherapy or even topical therapies such as cryotherapy (for BCCs), Efudix (topical fluorouracil), or Aldara (topical imiquimod), with the latter only appropriate for superficial lesions.\textsuperscript{[7]}

Here we report a case of very large, long-standing and neglected SCC of the scalp, which remained localised despite invading through the calvarium into the brain.

**Case presentation**

A 70-year-old previously well male presented with a large fungating mass covering the majority of the top of his cranium. History revealed this mass to be a recurrent SCC. Notably, the destructiveness of the lesion exposed bare skull as well as pulsating dura (Figure 1 and Video 1). There were areas of necrosis apparent throughout the large wound, however the extent of involved tissues was initially unclear. The lesion was not painful and the patient’s only complaint was of inconvenience and aesthetic appearance, describing having to cover it with a handkerchief and hat each day. It was evident that this lesion had developed through many years of neglect.

Due to his infrequent medical care, the patient presented with two other comorbid presentations, peripheral vascular disease and a large pelvic mass of size similar to that of a term pregnancy. While of concern, his scalp lesion was deemed more urgent and management focussed on resolution of this.

He was admitted into hospital for the management of his extensive scalp lesion. Preliminary wound cultures indicated a mixed organism infection and he was initiated on broad-spectrum intravenous antibiotics. Following 20 days of antibiotic therapy, the patient was stable enough for surgery and underwent craniotomy and excision of the lesion. Intra-operatively, it was found that the lesion had infiltrated through the dura and involved the brain parenchyma. Due to sagittal sinus bridging veins, only incomplete excision was possible. Reconstruction of the absent calvarium included the use of a latissimus dorsi flap with arterial and venous anastomosis. Histopathology confirmed moderately differentiated keratinising SCC with full thickness involvement of the dura.

Initial post-operative neurological signs were normal, with movement of all four limbs despite being externally ventilated. However, in the following days a mild left hemiparesis developed. The patient also became febrile, and a wound swab of the now partially necrotic muscle graft revealed growth of Proteus species bacteria. The patient returned to theatre for debridement of the necrotic tissue, however upon retraction of the flap, it was evident that the necrosis and infection was now involving brain tissue. The patient was returned to the intensive care unit and with consultation with family, the decision was made to withdraw ventilation and therapy. The patient died shortly afterwards.

**Discussion**

Invasion of NMSC into the brain parenchyma is extremely rare. There are very few such published cases and even less involve SCC, with the largest single case series published involving 26 patients treated at a single skull base centre of excellence over 12 years.\textsuperscript{[8]} These include Marjolin’s ulcers, in which the neoplasm develops within a chronic traumatic scar, typically involving a burn. The reasons for such aggressive local invasion without metastasis and the most appropriate management are largely unknown.

Previously deemed largely unmanageable, current treatment of extensive scalp SCC involves assessment by a multi-disciplinary team, including neurosurgical and plastic surgical input.\textsuperscript{[8,9]} Extensive wide excision including a dural excision is generally considered acceptable due to the resistance of the dura to tumour penetration. Following excision, reconstruction of all affected structures is necessary, using materials such as the fascia lata or a bovine pericardium graft (for replacement of the dura), titanium mesh (for replacement of the calvarium) and a musculocutaneous latissimus dorsi graft.\textsuperscript{[8]} This thorough, multidisciplinary approach has lead to up to 77.8\% local control rates at two-year follow up and some reports of disease control up to seven years following such surgery.\textsuperscript{[8,9]}

This approach does have its limitations and there are situations in which it is inappropriate. The dura is punctured by bridging veins, which drain the brain parenchyma into venous sinuses. Surgical disruption of these vessels in critical anatomical distributions can result in high mortality rates.\textsuperscript{[8]} Our case involved tumour spread to bridging veins in the distribution of the superior sagittal sinus, which resulted in incomplete excision. It is also important as to whether the tumour invades brain parenchyma, another situation presented
in this case. Resection margins of involved tissue is controversial and the extent of resection required to achieve appropriate clearance should guide decision-making. However, a large study from 2005 found that brain parenchymal involvement was statistically significant in decreasing disease-free survival regardless of surgical resection. Thus, in both of these scenarios, surgical intervention presents significant risks which when avoided, result in inadequate management.

Further, such surgery is highly invasive and given the predisposition for presentation of the elderly, the patient’s comorbidities and functional state are key predictors of post-operative morbidity and mortality. In our case, the concurrent identification of peripheral vascular disease, a large pelvic mass of unknown origin and culture-proven wound infection are significant considerations. The operative risk was clearly very large for this patient and these risks did manifest with the post-operative infection of his brain parenchyma and eventual death.

Defining the most appropriate management option in cases such as this is difficult and ultimately unclear. The first consideration is whether intervention is appropriate. In some elderly patients with multiple co-morbidities, intervention may be deemed a risk to quality of life. In certain circumstances, this is not an unreasonable approach and following discussion with the patient and family, could be considered. Should intervention be warranted but a patient’s operative risk too high, radiotherapy can be a useful alternative.

Radiotherapy is an underused but effective treatment option that can be considered as a definitive or adjuvant therapy in locally advanced SCC. There is much evidence that radiotherapy can produce similar rates of disease control and local regression when compared with surgery in early disease, and durable control rates in advanced disease. This efficacy, combined with a significantly lower procedural risk justifies its use as a definitive therapy. This is especially true in clinical scenarios such as head and neck lesions, where surgery is difficult and good cosmetic and functional outcomes are paramount to quality of life. Further, incompletely excised SCC has a high recurrence rate and up to 45% of recurrences involve regional metastasis. Should a patient be deemed appropriate for surgery, adjuvant radiotherapy can reduce the risk of local and regional recurrence and thus improve prognosis.

Our case illustrates how long a patient may survive with exposed brain tissue. The risk of sepsis or meningitis was obviously quite significant, yet was avoided with no particular wound care and generally poor wound hygiene practices. Given the asymptomatic mixed organism culture pre-operatively, it is possible that inadequate pre-operative eradication and subsequent surgical closure and containment of the infected tissues resulted in the fatal post-operative infection. The reasons why this infection remained asymptomatic for so long are not known, however regardless of this it seems likely that deterioration was inevitable in the near future.

Conclusion
This case presents a rare and difficult to manage locally advanced scalp SCC involving the brain parenchyma. Such cases generally occur in elderly people who often have poor general health and limited prognoses. While surgery is an appropriate management option, co-morbidities and other factors may increase the chance of post-operative complications or incomplete excision. Radiotherapy alone or in combination with surgery should be considered in the management of such cases. Alternatively, given the pre-operative stability of this patient and potential for decline with extensive neurosurgery, it may have been appropriate to consider withholding all interventions.

Conflict of interest
The authors declare that they have no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References
