

A Rare Neglected Pilomatrix Carcinoma on the Forehead: A Case Report

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ABSTRACT

Pilomatrix Carcinoma (PC) is seldom reported in the literature. These cancerous lesions present secondary to malignancy of the hair matrix. The consensus is that these malignancies tend to demonstrate an aggressive infiltrative pattern even in the face of local excision, with a subsequently elevated propensity for recurrence. Lymph node involvement and distant metastases are periodically witnessed. The primary treatment modality should comprise of surgery with adjuvant radiotherapy were deemed prudent. Regular follow up appointments are judicious since systemic metastases and nodal involvement can, unfortunately, unveil following the initial diagnosis and treatment plan. The case presented here illuminates an unusual encounter of a large cutaneous forehead pilomatrix carcinoma invading the frontal sinus, eroding the skull, and infiltrating the intradural/intracerebral compartment of the brain. This tumor was treated with wide local excision of the cutaneous tumor invading the surrounding skull, nasal bone with intradural/intracerebral components, en block excision of the bifrontal abscesses, and reconstruction with a scalp rotational flap and a split skin thickness graft harvested from the thigh for the cutaneous defect. The patient also had adjuvant radiotherapy as there was histological evidence of microscopic tumor cells at the margins of the resection. The patient has been followed up for 3 years and there were no signs of local or regional recurrence.

Keywords: Carcinoma; Pilomatrix; Cancer; Skin

INTRODUCTION

Pilomatrix carcinoma is purported to arise secondary to malignant transformation of the benign cutaneous pilomatrixoma [1]. They can commonly present in the clinic as an asymptomatic, indurated solid or cystic lesion varying from around 0.5 cm-20 cm. Diagnosis is based on the histological examination of biopsied lesions. The benign pilomatrixoma is commonly identified in the younger cohort between the ages of 0-20, especially in the head and neck region [2,3]. Pilomatrix carcinomas most commonly occur in males at around the age of 45 years. Literature searches using high evidence sources such as PubMed and Embase databases yielded fifty-one cases of a pilomatrix carcinoma presenting in the head and neck region. With an insufficient range and scantiness of cases reported, treatment planning can be challenging and requires the meticulous utilization of a Multidisciplinary Team (MDT) entailing a maxillofacial surgeon, neurosurgeon, oncologist,

histopathologist and dedicated specialist head and neck nurses. With a paucity of case reports and literature on this topic, we hope to bestow evidence of good clinical practice and management for a rather scarce type of neoplasm.

CASE REPORT

An 80-year-old gentleman presented to the accident and emergency department with a long history of an neglected mass on his forehead and a subsequent bifrontal sinus abscess culminating in marked cognitive impairment. His Glasgow Coma Scale was 8 and he was clinically aphasic and had marked difficulty mobilising. He was rapidly deteriorating in the emergency department and an urgent CT head was ordered which showed an aggressive lesion in the forehead area eroding at the frontal bone and invading the cerebral cortex. The lesion measured 48 × 73 × 51 mm. At a later date, an MRI with contrast was conducted with both special investigations

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confirming an aggressive tumor invading the frontal sinus, frontal bone, cribriform plate, and extending intracranially. Clinically and dermoscopically, the lesion appeared to resemble a basal cell carcinoma. Prior to surgery, the patient had a wedge biopsy which revealed a basal cell carcinoma with hair matrix differentiation (resembling a pilomatrixoma carcinoma). Following the multi-disciplinary team meeting, the patient was taken to theatres whereby en block excision of the bilateral frontal abscess was undertaken by the neurosurgeons and wide local surgical excision of the macroscopic component of the tumor by the maxillofacial team. Sections of the frontal bone were also removed alongside the external components of the meninges-primarily the dura matter. The surgical site was closed with a craniofacial rotational flap and a split skin thickness graft harvested from the left thigh. There were no complications in recovery or the days following the surgery on the ward. There was a large bony defect created secondary to the partial craniectomy which was planned for a secondary reconstruction following the adjuvant localized radiotherapy-this never materialized as the patient was content with the outcome and the deformity did not affect his quality of life [4]. Staging of the cancer was completed via a combination of an ultrasound of the neck and computed tomography of the chest and abdomen which was negative for metastatic disease. He received 60 Gy radiotherapy in 30 daily fractions over six weeks using 6 MV photons and a rapidArc technique with bolus into the craniectomy defect. In terms of toxicities, the patient suffered from grade 1 fatigue [5], hyperlacrimation with no evidence of conjunctivitis, and brisk erythema grade 2a radiation oncology/toxicity grading alongside dysgeusia [6]. These complications gradually settled over 6 months 3 years post-surgery, the latest MRI Head with contrast revealed no evidence of local or regional recurrence and thus no need for any further surgical intervention.

Histopathology

The microscopic analysis of the tumor revealed 'basal cells forming islands with typical peripheral palisading of nuclei and clefting. There are many mitoses and apoptotic bodies. However, the most unusual feature is squamous differentiation with host cell epithelium, a foreign body giant cell reaction, and a fibroblastic reaction with mixed acute on chronic inflammation. Perineural invasion was also seen. This appearance resembles a pilomatrix carcinoma. With regards to the tissue removed from the frontal sinus, the conclusion was of 'acute or chronic abscess/empyema with foreign body reaction to non-viable matrix ghost cell epithelium'. As communicated earlier, the neoplasm did extend to the margins.

DISCUSSION

Pilomatrix Carcinoma (PC) is an uncommon malignant skin adnexal neoplasm derived from the hair matrix [7]. De Novo formation or mutation from a benign pre-existing Pilomatrixoma tends to remain a contentious topic in the literature [8]. What is more fascinating is that I could not find a single case of a pilomatrix carcinoma on the forehead. It has been postulated that the formation of both benign and malignant pilomatrixoma is due to mutations in exon 3 of the

beta-catenin gene [9]. The lesion can follow an indolent course with the diagnostic predicament in being competent to discriminate the histology of this lesion to similar neoplasms such as pilomatrixoma, proliferating pilomatrixoma, and a basal cell carcinoma [10]. Other differential diagnoses for this tumor can include malignant melanoma, Merkel cell carcinoma, squamous cell carcinoma, and vascular lesions [11]. From a histopathology perspective, the key to discriminating between the proliferating pilomatrixoma and pilomatrix carcinoma lies in the fact that there are more prominent perineural/vascular invasions, nuclear atypia and atypical mitoses present in the Pilomatrix carcinoma [11]. In terms of epidemiology, PCs tend to have a predilection for a certain cohort of the population with white middle-aged men twice as likely to procure this malignant disease [10,12]. Unfortunately, there are no known risk factors for the formation of these tumors, and around 60% of these cases present in the head and neck region [13].

In terms of the histology, these malignancies tend to demonstrate central areas of necrosis, clusters of anaplastic cells, hyperchromatic basaloid cells, nuclear pleomorphism, the transition to squamous cells, lymphatic infiltration alongside an infiltrative growth pattern [14,15]. In terms of remission, studies have indicated that a recurrence-free interval of 5-17 months following the first resection can be observed, whereas other studies have seen evidence of local scalp recurrence 6 months after resection [10,16]. Local tumor recurrence can also be evidenced despite the presence of clear surgical margins [10]. Combining all the currently available evidence on this neoplasm, it has been identified that with wide local excision, a recurrence rate of 23% can be seen at 7 months or later compared with an 83% recurrence of simply excised tumors at an average of 11.9 months or later following the primary excision [17]. From a statistical perspective, this equates to a 53% recurrence rate [16,17]. Given these statistics, it would seem prudent that wide local excision should be exercised and implemented in all subsequent pilomatrix carcinomas. It is, however, also accepted that wide local excision can be omitted due to the presence of certain anatomical features, such as the frontal lobe of the cerebral cortex in this case-invasion of which could lead to devastating consequences for the patient's subsequent quality of life.

Whilst the definition of adequate surgical margins is a controversial subject, reports of margins between 5 mm-2 cm have been widely cited as sufficient amongst different surgeons [18]. An alternative to wide local excision could include the conservative Mohs micrographic surgery to ensure margins are devoid of tumor cells and thus guaranteeing comprehensive histological margin control [19]. If feasible, nodal metastasis should be treated with the relevant nodal dissection and the possibility of a sentinel lymph node biopsy should also be deliberated. Of note is that in over 50% of patients presenting with nodal metastasis, coexisting systematic metastatic disease is also present. As a general guide, 13% of the reported cases in the literature have presented with lymph node and systemic metastatic disease [17]. As in most oncology cases, the primary mechanism driving the prognosis of cancer is systematic metastases where if present a collective decision by the MDT should be made to ascertain whether the patient is fit for

surgery or palliative care [20]. The role of radiotherapy is yet to be explicated, but the prescription of adjuvant radiotherapy following excision has been deemed to provide acceptable levels of local tumor control [21]. In addition to this, radiotherapy could also be used at the discretion of the oncologist for palliative management of systemic metastatic disease. With regards to chemotherapy, it is reported that systemic disease is unresponsive to traditional chemotherapeutic agents and its administration in patients implicated with systemic metastatic disease secondary to PC is somewhat futile [22,23].

CONCLUSION

In conclusion, it is imperative to be competent in correctly diagnosing a pilomatrixcarcinoma given its somewhat surreptitious ability to spread systemically as appose to the generally more indolent basal cell carcinoma. It, therefore, should form part of the differential diagnoses when indurated tumors of the head and neck region are encountered. In terms of treatment, priority should be given to wide local excision where feasible alongside adjuvant radiotherapy where there is speculation regarding the uncertainty of the tumor-free clearance of the histological margins. As systemic metastases are rarely encountered at initial diagnosis, follow up examinations should be considered mandatory in ensuring early detection of any recurrence or evidence of nodal/systematic metastases.

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CONFLICT OF INTEREST

The authors declare that there are no conflicts of interest.

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