A Persistent Case of Periocular Cystic Squamous Cell Carcinoma

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Abstract

A 53-year-old man with a history of multiple cutaneous facial squamous cell carcinoma (SCC) presents with a progressively enlarging cystic mass of his right brow for two months associated with periocular pain and numbness along the right side of his scalp. In addition, several subcutaneous lesions were noticed along the nasolabial and nasojugal areas. All lesions were excised and confirmed to be SCC on pathology with perineural spread observed with the primary brow lesion. The presence of a cystic lesion and/or multiple subcutaneous lesions should prompt consideration of a metastatic presentation of SCC. Furthermore, perineural invasion of SCC can be associated with sensory and/or motor deficits.

Key Words: squamous cell carcinoma, periocular, cyst, perineural invasion, subcutaneous metastasis.

Introduction

Squamous cell carcinoma (SCC) is the second most common eyelid malignancy in Caucasians and accounts for less than 5% of malignant eyelid neoplasms.¹ SCC is a neurotrophic tumor and has the aggressive potential to become metastatic through perineural spread (PNS) often manifesting with facial dysesthesias and/or motor nerve palsies in the absence of visible masses or as subcutaneous metastasis appearing as satellite lesions.² ⁴ Although rare, there have been reports in the literature of PNS of SCC associated with a cystic lesion,³ however this case describes recurrence of SCC with a cystic component and associated PNS, as well as, the development of atypical lesions in the subcutaneous tissue likely representing metastatic spread and advanced disease.

Case Report

A 53-year-old man presents with complaints of pain and right-sided facial numbness along with a mass of his right upper eyelid. The mass had been present without symptoms for two months but recently became painful and swollen. The medical history was significant for excision of several squamous cell carcinomas of his face leaving a large surgical defect of the right maxillary sinus and his entire nose. On examination there was tender, cystic and mobile mass measured at 4.0 x 2.0 cm, located anterior to the superior orbital rim of the right eyebrow (Figure 1a). Several subcutaneous lesions were also noticed: two in the left nasolabial (3.5 x 3.0 cm and 2.5 x 2.0 cm), one on the right nasolabial (4.3 x 3.0 cm), one in the left nasojugal (3.0 x 1.0 cm) and one eroding the medial orbital and anterior lacrimal crest, extending towards the brow (2.5 x 2.5 cm) (Figure 1b). A right lower lid cicatricial entropion was present.

Figure 1: (a) Clinical photograph showing a large cystic SCC of the right brow and (b) multiple subcutaneous lesions along the nasolabial and nasojugal areas prominent on the left side of his face.
along with paraesthesia along the V1 and V2 dermatome on the right. Best corrected visual was 20/25 OU with remainder of his ocular examination, including slit-lamp and dilated ophthalmoscopic examination was normal. A right medial orbital infiltration with opacities of the right ethmoidal sinuses was seen on CT.

The patient underwent a right brow tumor excision along with excision of the other facial subcutaneous lesions. Histopathologic examination was completed with Hematoxylin & Eosin and immunoperoxidase stains including both S100 protein for identifying nerves and MNF116 as a keratin marker (Figure 2a-2b). The brow lesion revealed poorly differentiated SCC with perineural invasion along the frontal nerve explaining the hypoesthesia. Examination of the subcutaneous lesions demonstrated malignant squamous epithelial tumor deposits and foci of necrotic SCC, as well as, scattered small nerve branches within the specimen, showing perineural and occasionally intraneural invasion.

Based on his previous history, metastatic SCC was the most likely the diagnosis. The surgical margins were positive with evidence of perineural spread in the primary brow lid lesion and metastatic disease was seen in the subcutaneous lesions. He has received maximal radiotherapy and has declined exenteration; unfortunately, our patient prognosis was poor. Management is now aimed at palliative care for pain and control of symptoms of this advancing disease.

**Discussion**

PNS associated with SCC of the head and neck may present in a variety of settings and cause significant morbidity and mortality. The confirmation of PNS despite suggestive signs and symptoms can prove difficult and it maybe until disease develops within the orbit, cavernous sinus and/or nerves/foramina that a diagnosis can be supported. PNS can present clinically in patients who complain of periorcular pain, dysesthesias or paraesthesia along the trigeminal dermatome and/or ocular motility deficits. If a patient presents with pain and periorcular and ocular neurological symptoms the diagnosis should be suggested even if the cutaneous lesion cannot be identified.

The pathophysiology of PNS associated with cystic SCC has not been well characterized. The presence of subcutaneous nodules along the pathway of the effected nerve has been noted and suggests that SCC penetrates beyond the nerve sheath. The microscopic appearances suggest that the tumor extends along the perineural space and penetrates through the epineurium to gain access to the orbital connective tissue. Initially the lesion appears to maintain a solid structure, but with persistent growth there is progression to central necrosis and secondary cystic degeneration that leaves a thin rim of viable neoplastic epithelium lining the structure. The presentation of cystic SCC has been described on imaging (CT and MRI) and linked to PNS. The cases with radiographic evidence of PNS has been shown to be associated with a poorer prognosis (50% 5-year survival rate) than imaging-negative disease (86% 5-year survival rate).

The occurrence of SCC in the secondary lesions excised likely demonstrated subcutaneous metastases. Clinical studies suggest that the cutaneous spread of SCC is caused by lymphatic dissemination and/or PNS and if distant possibly hematogenous spread. Metastases are often site-specific to areas of optimal cell proliferation and tend to be close to the primary tumor and areas of lymph node spread. This is slightly different in our case in that most of the subcutaneous lesions on presentation were located on the left side, whereas excisions of previous SCC were on the right side of the face. Studies have shown that subcutaneous spread of SCC in the head and neck have a poor prognosis.

Treatment recommendations for PNS and/or dermal metastases of SCC vary and investigations determining the extent of disease will guide treatment. There is good evidence that combined modality treatment with surgery and adjuvant radiotherapy improves locoregional control.
and survival in head and neck SCC. Surgery serves mainly for diagnostic purposes, as complete removal of an extensive lesion is unrealistic. It is likely in this case surgery can also be performed for palliative care as debulking the tumor can manage pain and minimize symptoms. In the setting of aggressive microscopic spread of disease, radiotherapy targeting the lesion and the possible areas of spread is the mainstay of treatment. Radiotherapy can be curative with early disease, but in advanced disease, the aim is growth restraint, reduction in local morbidity, and pain control. Despite extensive surgery and significant radiation therapy our patient underwent, the tumor continued to grow and invade the surrounding structures of his face. All treatment is now focused on palliative care.

In conclusion, the presence of a cystic lesion in a patient with a history of SCC of the face and ocular adnexa or sensory and/or motor deficits of the face in the periorcular area is suggestive of PNS from a SCC. Further suspicion for metastatic disease should arise if small subcutaneous lesions arise by the primary site of malignancy. These characteristics highlight advance disease and should prompt for urgent biopsy and possible imaging studies to determine the extent of the disease and establish treatment.

REFERENCES