Case Report

DEVIL IN DISGUISE: BASOSQUAMOUS CARCINOMA OF EYELID

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ABSTRACT

Basosquamous carcinoma is a rare epithelial malignancy of the eyelid with aggressive and destructive behaviour. They share the features of both squamous cell and basal cell carcinoma. We report a case of a patient who presented with left lower lid enlarging mole and clinical signs of basal cell carcinoma . There were local metastases to the orbit and paranasal sinus. Histopathology revealed infiltration of basaloid neoplastic epithelial cells into underlying dermis in cords and strands with peripheral palisading and stromal proliferation. Patient underwent exenteration of the affected eye along with radiotherapy. There has been no recurrence of tumour at 8 months post radiotherapy.

INTRODUCTION

Eyelid tumours are by far the most common neoplasms encountered in clinical ophthalmic practice [1]. Approximately , 90% of skin cancers arise in the head and neck region and 10% of them occur in the eyelid [2]. Basal cell carcinoma is the most common eyelid tumour and constitutes 80 - 90% of its bulk [3]. The second commonest eyelid tumour is the squamous cell carcinoma which forms 5-10 % of the eyelid malignancy [4]. Basal cell carcinoma has little metastatic potential and squamous cell carcinoma has a reported metastatic rate up to 21% [5].

Basosquamous carcinoma (BSC) also known as metatypical carcinoma, basaloid squamous cell carcinoma or keratolytic basal cell carcinoma. They share the features of both squamous cell and basal cell carcinoma. Basosquamous carcinoma is an extremely rare epithelial malignancy of the eyelid with aggressive behaviour. They form less than 2% of incidence of all non-melanomas of the skin [6]. This malignant neoplasm is now considered a new variety of non-melanoma skin cancer with its own characteristics and histologic characteristics. In some cases, distinction between a basal cell and squamous cell carcinoma can be difficult and definitive identification of a BSC may be more challenging [7]. We are reporting a case report of a patient with presentation of BSC.

CASE REPORT

A 53-year-old gentleman who was previously healthy, presented with enlarging mole of the left lower lid for the past 5 years. The lesion had grown progressively larger and became more violaceous with on and off contact bleeding in the past one year. Patient sought medical attention after 5 years of presentation when he developed left mechanical ptosis. The best corrected visual acuity (BCVA) was 6/7.5 in the right eye and hand movement in the left eye. Left ocular examination revealed an ulcerated and hyperpigmented mass over both upper and lower lid region with contact bleeding. The mass was hard and indurated associated with mechanical ptosis (Figure 1A). The bulbar and palpebral conjunctiva were congested with feeder vessels seen (Figure 1B). There was left eye relative afferent pupillary defect (RAPD). Anterior segment and fundus examination were normal. Examination of the right eye was unremarkable. There was no cervical lymphadenopathy.

A computed tomography (CT) scan of the orbit and brain revealed heterogeneously enhancing soft tissue mass in the lower, lateral, and posterior aspect of the left orbit. There was also involvement of left inferior rectus and left inferior oblique muscles with extension of mass to the anterior aspect of optic nerve. The mass extends superficially to lower

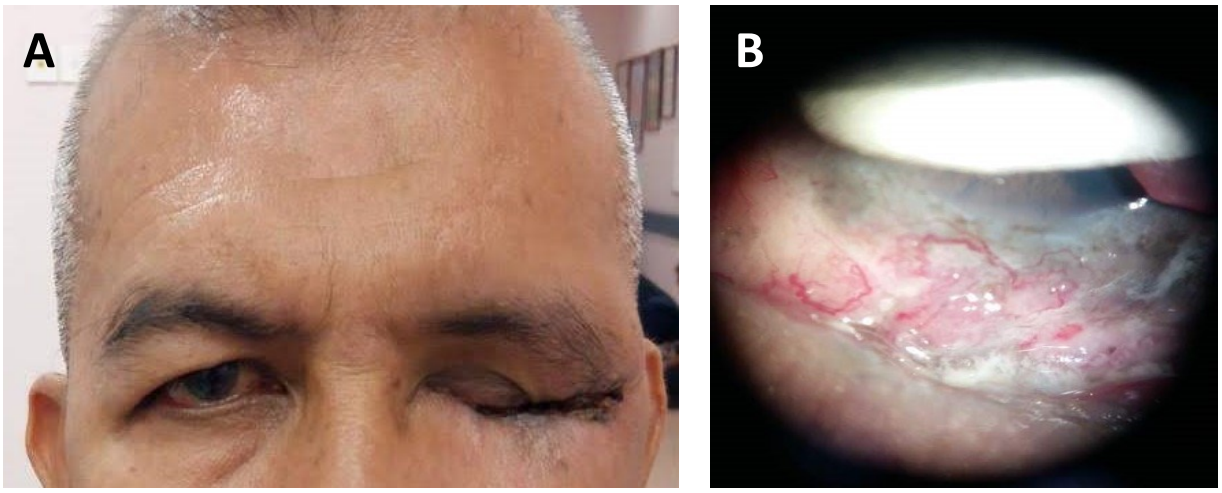

Figure 1A: Left mechanical ptosis with hyperpigmented and ulcerated lesion over the upper and lower lid.
Figure 1B: Left palpebral conjunctiva is congested with feeder vessels seen.

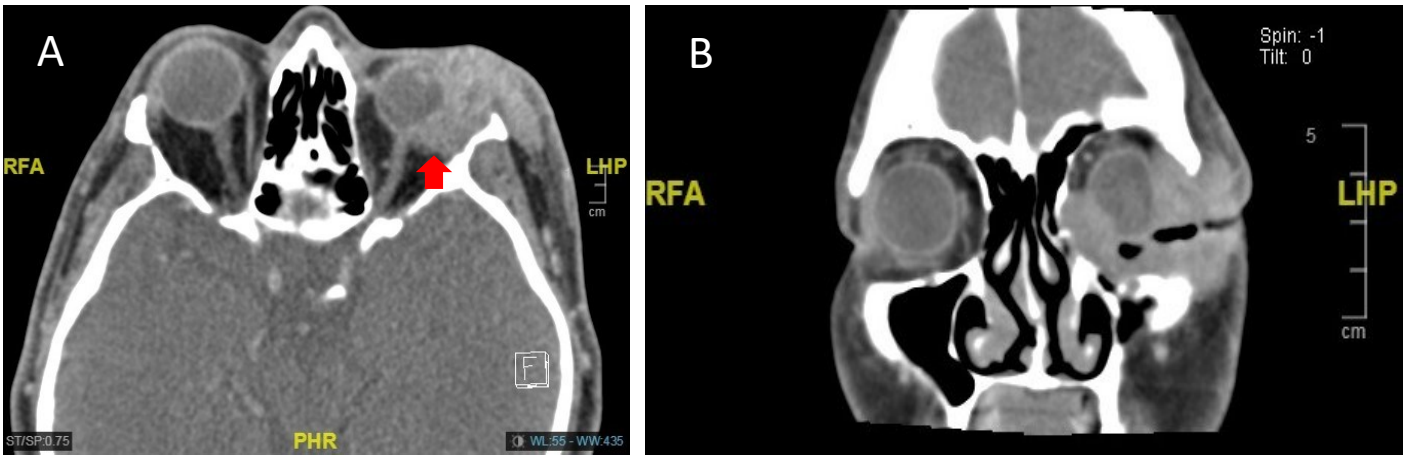

Figure 2A: Computed tomography (CT) scan of the orbit with axial view showing irregular heterogenous mass filling the intra and extraconal spaces within the left orbit (red arrow). Half of left lateral, medial and inferior recti muscles is involved with loss of the normal anatomical configuration and ocular space, causing deformed left globe.

Figure 2B: There is an extension of the mass to the anterior aspect of optic nerve. CT scan of orbit with coronal view showing mass extends superolaterally to left lacrimal gland, laterally to left zygoma and inferiorly into left maxillary sinus with associated bony destructions.

eye lid, superolaterally to left lacrimal gland, laterally to left zygoma and inferiorly into left maxillary sinus associated with bony destructions (Figure 2).

Subsequently, incisional biopsy was done. Histopathological examination showed infiltration of basaloid neoplastic epithelial cells into underlying dermis in cords and strands, forming reticulated pattern. The neoplastic cells showed peripheral palisading, and the surrounding stroma was desmoplastic with occasional mucin deposition seen. Tumour clefting from the stroma was observed as well. Immunohistochemical stains showed the neoplastic cells were diffusely positive for Epithelial antigen (Ber-EP4) and negative for Epithelial membrane antigen (EMA). Thereby, a histopathological diagnosis of left basal cell carcinoma was made.

In view that patient had left basal cell carcinoma with extensive extension into orbital and paranasal sinuses, patient was counselled for left extended exenteration and the surgery was proceeded (Figure 3A). Histopathological examination of the exenterated eye and paranasal sinus showed that the tumour was composed of islands, solid nests, cohesive sheets, and clusters of closely packed basaloid cells with round to oval nuclei and minimal cytoplasm. Foci of squamous differentiation were found. The tumour cells were Ber-EP4 positive. Areas of tumour necrosis and perineural invasion were also seen. Histopathological diagnosis of left eyelid BSC with orbital soft tissue and bone invasion was made.

A CT scan of the thorax, abdomen and pelvis revealed no metastases. At one month post

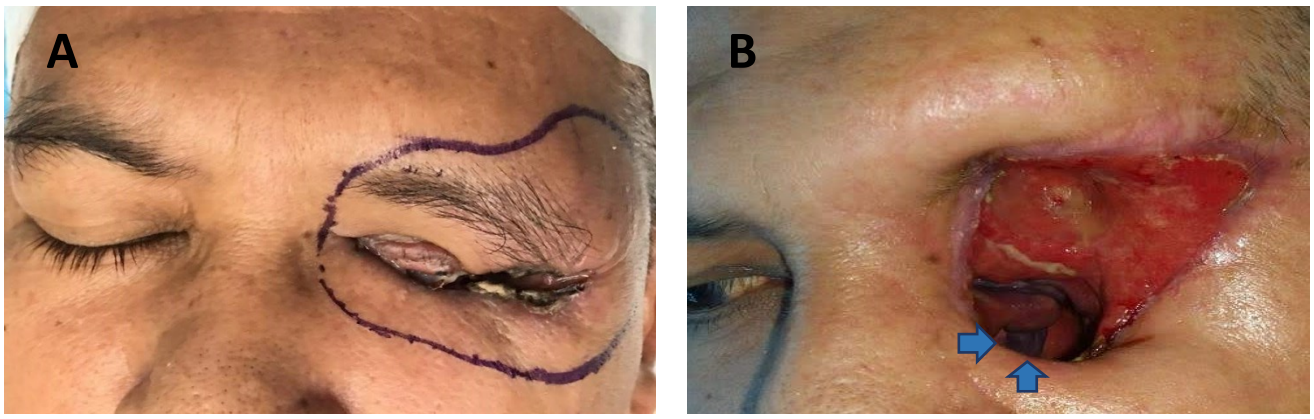

Figure 3A: Left eye mechanical ptosis with hyperpigmented and ulcerated mass causing destruction of eyelid architecture. Surgical excision area marked prior to exenteration.
Figure 3B: At one month post exenteration: the wound and socket is clean with healthy granulation tissue. Blue arrows indicate the maxillary sinus.

exenteration, the wound was clean and healthy granulation tissue observed (Figure 3B). Patient subsequently underwent radiotherapy. There was no recurrence noted at post radiotherapy.

DISCUSSION

Basosquamous carcinoma was first described by Mac Cormac in 1910 as a rare tumour that formed less than 2% of the incidence of non-melanoma skin cancers. This histologic variant was reported in a series of rodent ulcer in which both basal cell and squamous cell carcinoma was present without any transition zone [8]. In 1974, the World Health Organisation confirmed BSC as a separate entity [9]. Previous studies have not provided the exact definition of BSC except for Burston and Clay. According to Burston and Clay, BSC is defined as BCC differentiating into squamous cell carcinoma [10].

Basosquamous carcinoma is a slow growing neoplasm but highly aggressive. There is a wide variation of clinical presentation of BSC as in our case report, which makes it difficult to differentiate BSC from squamous cell carcinoma.

Clinically BSC is indistinguishable from the basal cell carcinoma. Histologically, BSC is defined as a neoplasm with the features of basal cell carcinoma with foci of squamous differentiation and spindle cell ar-eas. The tumour lobules are more irregular and pe-ripheral palisading is less pronounced but focally present. Stromal proliferation is more prominent, as presented in our case [11].

Immunohistochemistry also aids in the diagnosis of BSC. Basal cell and squamous cell carcinoma can be readily distinguished using routine immunohisto-chemistry for Ber-EP4 and EMA [12]. Basosquamous carcinoma BSC showed an immunostaining pattern similar to that of basal cell carcinomas with Ber-EP4 positivity and with negative staining for EMA, similar to our patient. Using the help of the immunohistochemistry, it could help in diagnostic certainty of this rare challenging BSC.

The treatment modalities for BSC are surgical excision, radiotherapy, and Mohs microscopic surgery [13]. Surgical excision is the treatment of choice. For BSC, surgical margins should be wider than those for low-risk basal cell carcinoma due to the infiltrative growth pattern of this tumour. However, despite a wide local excision, there has been high rate of recurrences reported. According to Borel et al, he reported a local disease recurrence rate of 45.7% after wide excision for BSC in 35 patients with a follow-up period of 1 year [14]. Another study done by Schuller et al, reported that a recurrence rate of 12.1% in 33 patients with BSC treated with surgical excision [15].

Adverse prognostic signs include perineural and lymphatic spread, similarly observed in our case. According to Petri et al, patients with perineural invasion have significantly higher rates of metastatic disease and local recurrence compared to patients without perineural invasion [16]. Thereby, a close and a long clinical follow up is needed for this aggressive primary tumour.

CONCLUSION

In conclusion, BSC is a rare and aggressive form of carcinoma with propensity of lymph node and perineural spread. Most BSC arise in the head and neck, so in our case report, it provides important insights into this tumour entity. The rarity of this tumour, the pattern of growth, and the initial lack of diagnostic criteria have resulted in only a few studies evaluating the best treatment options for patients. Thereby, this tumour is rightly described as devil in disguise due to its aggressiveness and destructive behaviour.

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