

Peri - and Intraocular Mutilating Advanced Squamous Cell Carcinoma: "Monsters Inside Your Body"?

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Abstract

Periocular malignancies represent between 5% and 10% of all types of skin cancers. The incidence of eyelid (but also the periocular located) malignancies seems to differ in distribution across the continents. The incidence of eyelid tumours (but also the periocular located tumours) in a predominantly white population determined that BCC is the most common malignant periocular eyelid tumour in whites. This finding has been replicated consistently throughout the literature, with BCC representing 85–95% of all eyelid malignancies, SCC representing 3.4 - 12.6%, Seb Ca representing 0.6 - 10.2%, and both melanoma and Merkel cell carcinoma representing less than 1%. Most periocular skin cancers are associated with ultraviolet radiation (UVR) exposure. Ultraviolet radiation causes local immune suppression, which, coupled with DNA abnormalities in tumour suppressor genes and oncogenes, leads to the development of skin cancers. We are presenting a 62 - year - old patient with a small nodule about 2 cm away from the lower lid of his left eye. A tumour was surgically treated. Several years later there was a tumour relapse, treated with radiotherapy and subsequent chemotherapy with Endoxan and Cisplatin. After the second relapse, he was treated surgically in general anaesthesia by orbital exenteration, removal of the orbital floor and resection of zygomatic bone and the maxillary sinus. A couple of months later, he developed a tumour relapse in the scars and the area of a primary tumour with tumour progression. A possible therapy with Cetuximab or radiation therapy was discussed as a possible treatment option.

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Introduction

Periocular SCC is a tumour with a possible unfavourable outcome, when not treated efficiently primarily. A recent study from Australia investigated the recurrence of SCC according to TNM classification. They observed a recurrence rate of 5.3% (12/226 patients) for primary tumors and 20% (5/25 patients) for recurrent tumors (P = 0.019). Recurrences also occurred in T1 tumours. Higher T stage was significantly associated with both perineural invasion and local recurrence [1].

Perineural invasion capacity of periocular SCC bears a significant risk of orbital involvement and the need subsequent orbital exenteration [2]. There is a great variety of reconstructive procedures, but orbital prosthesis and simple split skin grafts may be helpful as well [3].

Adjuvant treatment with targeted tumour therapy is under investigation. Cetuximab has achieved a response rate of ≤ 47 %, erlotinib and gefitinib – to other epidermal growth factor tyrosine kinase inhibitors are under investigation in adjuvant and neoadjuvant settings [4][5][6].

Case presentation

A 62 - year - old male patient presented in 2009 with a small nodule about 2 cm away from the lower lid of his left eye. His medical history was positive for hypertension and unspecified hepatitis. The tumour was surgically removed elsewhere (Fig. 1 a, b).



Figure 1: a, b - Mutilating postoperative results and tumour relapse

It was a squamous cell carcinoma (SCC), but the resection was obviously not R0. There was a relapse, and the tumour developed rapidly until 2013. From 09.09.13 to 13.09.13 radiotherapy had been performed. In March 2014, he received a single course of systemic chemotherapy with endoxan and cisplatin because of a second relapse. Computerized tomography (CT) revealed extended osteolysis of left sinus maxillaries, nasal bone and orbital bone. The polypoid soft tissue process involved not only sinus maxillaries and overlying subcutaneous adipose tissue but ocular bulbus as well. Chemotherapy was stopped.

He was treated surgically in general anaesthesia by orbital exenteration, removal of the orbital floor and resection of zygomatic bone. The tumour resection included the maxillary sinus. The orbital defect was covered by temporal muscle. The soft tissue defect was closed by fronto-parietal rotational flap, buccal plasty, and a mesh graft obtained from his upper leg. A couple of months later, he developed a tumour relapse in the scars and the area of a primary tumour with tumour progress.

Discussion

The standard treatment for all eyelid carcinomas is surgical excision with negative margins, although controversy still exists regarding the recommended margins for each specific malignancy [7]. Mohs micrographic surgery has become the most common method of managing both BCC and SCC [8]. The benefits of this procedure lie in its capacity to determine margin control during excision and preserve the greatest amount of normal tissue [8]. This is especially significant in the eyelid and medial canthus, where large excisions can have devastating effects on the appearance and function of the eyelids [8]. Both frozen and permanent sectioning has been used for histologic assessment of surgical margins [8]. There does not seem to be a consensus in the literature regarding the preferred margin control technique for nonmelanoma malignancies [8].

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