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SHORT COMMUNICATION

Squamous cell carcinoma presenting as an orbital cyst with radiologic evidence of perineural invasion

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PURPOSE. *To report clinical and radiologic findings of cystic squamous cell carcinoma (SCC) of the orbit with evidence of perineural involvement.*
METHODS. *Analysis of clinical findings and radiology with a literature review.*
RESULTS. *A 66-year-old man with SCC of the forehead 8 years prior presented with paraesthesias, diplopia, and proptosis. Magnetic resonance imaging showed a well-defined, cystic mass of the orbit with a single, linear structure running through its center. Lateral orbitotomy revealed a cyst adherent to adjacent periorbita containing viscous, clear, yellow substance and a nerve coursing through the center. Histopathology confirmed poorly differentiated spindle cell carcinoma with positive staining for cytokeratin markers, consistent with SCC.*
CONCLUSIONS. *Orbital cysts associated with altered sensation are suggestive of SCC with perineural spread, requiring prompt investigation and treatment to minimize morbidity and mortality. The involved nerve may be seen as a single, linear structure within the mass on imaging. (Eur J Ophthalmol 2007; 17: 970-2)*

KEY WORDS. *Cystic orbital mass, Cystic squamous cell carcinoma, Orbital cyst, Orbital squamous cell carcinoma, Periocular squamous cell carcinoma*

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INTRODUCTION

Squamous cell carcinoma (SCC) is the second most common eyelid malignancy and its incidence is increasing. Unlike the more common basal cell carcinoma, SCC is a neurotrophic tumor and perineural infiltration in the periocular region may facilitate spread into the orbit and intracranial cavity, causing significant morbidity and mortality (1, 2). Rarely, it may present as an orbital cyst with evidence of the affected nerve on imaging (2, 3). The appearance of the affected nerve on imaging has not been well-described. We report a case of cystic SCC of the orbit with a linear intensity found within the mass on magnetic resonance imaging (MRI) which correlated with intraoperative findings and suggested tumor spread along the lacrimal nerve.

Case report

A 66-year-old man had 2 months of paraesthesias in the distribution of the right lacrimal nerve. He had a history of moderately differentiated invasive SCC of the right forehead 8 years prior, which was reported to be completely excised. On examination, he had diplopia in extreme upgaze and 2 mm of right proptosis. Computed tomography (CT) of the orbits demonstrated a well-defined, cystic

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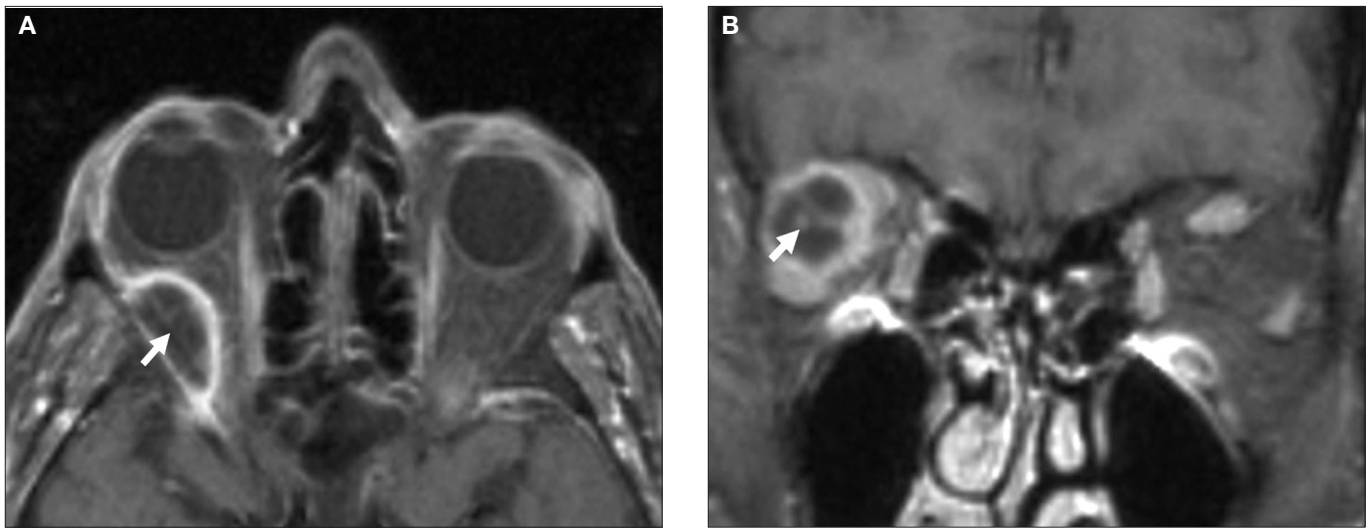

Fig. 1 - (**A**) *T1-weighted, contrast enhanced, fat suppressed, axial magnetic resonance image (MRI) of the orbits confirming a well-defined cystic mass. A linear hyperintensity was seen running within the mass, which given the history of paraesthesia, was presumed to be the lacrimal nerve (arrow).* (**B**) *T1-weighted, contrast enhanced, fat suppressed, coronal MRI of the orbits reveals a hyperintensity in the center of the mass, corresponding to a nerve seen within the cyst intraoperatively.*

retrobulbar mass in the superotemporal orbit. MRI of the orbits (Fig. 1) confirmed the cystic nature of the mass with enhancement of the rim of the lesion. In addition, a linear hyperintensity running through the center of the cyst could be seen. Lateral orbitotomy revealed a cyst lining adherent to adjacent periorbita, orbital fat, and lateral rectus muscle. A viscous, clear, yellow-tinged substance was present within the mass. Furthermore, a nerve was found coursing through its center which correlated with the linear hyperintensity on MRI. The bulk of the cyst was carefully excised; however, residual adherent tissue remained. Histopathology showed poorly differentiated spindle cell carcinoma with positive staining for cytokeratin markers, consistent with SCC (Fig. 2). The cytokeratin markers which stained strongly positive included AE 1/3 (Dako), Callus keratin (Dako), Bovine keratin (Dako), and CAM 5.2 (Becton-Dickinson). In addition, there was no evidence of an underlying dermoid cyst. Examination of his skin and further investigation including CT of the neck, chest, and abdomen failed to reveal a primary tumor or disseminated disease. The patient refused postoperative radiotherapy due to the risk of vision loss.

Within 4 months, he developed orbital recurrence and underwent exenteration with free microvascular flap reconstruction, followed by radiotherapy. Six months after exenteration, MRI showed no evidence of disease progression.

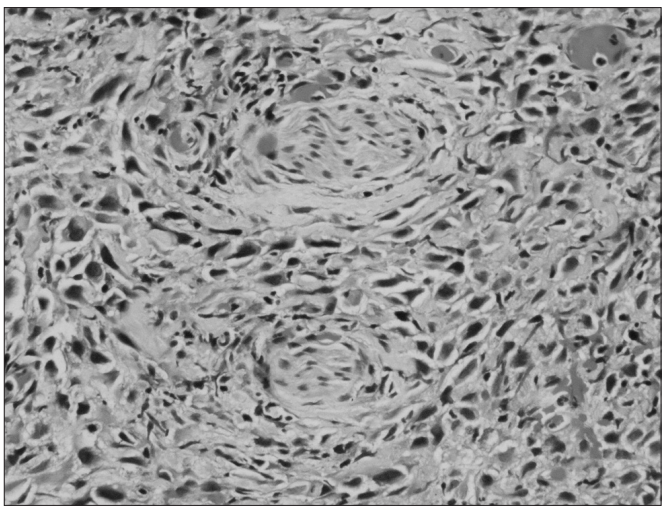

Fig. 2 - *Histopathology showed poorly differentiated spindle cell carcinoma with perineural invasion (HEX40). Stains were positive for cytokeratin markers, consistent with squamous cell carcinoma.*

DISCUSSION

Perineural invasion (PNI) describes tumor growth in or around a nerve and is confirmed on histopathology in 4 to 8% of periocular SCC (4, 5). It is associated with large (>2 cm), head and neck, previously recurrent, and poorly differentiated SCC. Cutaneous SCC may spread to the orbit by perineural invasion, commonly presenting with numb-

ness, pain or formication in the distribution of the first and second branches of the trigeminal nerve, and motor nerve palsies (1, 3). SCC has been reported in association with cystic choristomas of the orbit; however, these lesions may have actually resulted from perineural spread as this was confirmed on histopathology in one case (6, 7). In a study of 17 patients with involvement of the orbit and ocular adnexa by distant perineural spread of SCC, 5 had evidence of a cystic lesion on imaging (1). The involved nerve may be seen on imaging; however, this has not been well-characterized in prior studies. This report illustrates that SCC of the orbit may rarely present as a cystic mass and hence should be considered in the differential diagnosis of an orbital cyst. Furthermore, a linear hyperintensity coursing within the cyst may be seen on imaging, suggesting nerve involvement. Ophthalmologists should be alerted that any periorbital cystic lesion associated with altered sensation may contain a characteristic single linear density on imaging and is suspicious for SCC with PNI, requiring prompt investigation and treatment to minimize morbidity and mortality.

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