



CASE REPORT

Metastatic Squamous Spindle Cell Carcinoma of the conjunctiva

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KEYWORDS

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Abstract Conjunctival Squamous Cell Carcinoma (CSCC) is one of the important ocular surface tumors. It is thought to be more advanced in this part of the world such as in Saudi Arabia. The Squamous Spindle Cell Carcinoma (SSCC) is a more aggressive variant. A single case of spindle squamous cell carcinoma tumor-related death due to bone and lung metastasis has been reported in Sweden in 1995.

We are reporting a similar case of SSCC of the conjunctiva with eventual death because of bilateral lung metastasis in our area. Proper identification of this type of tumor by ocular pathologists and the need for future detailed clinicopathologic studies on CSCC in Saudi Arabia is emphasized.

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1. Introduction

Conjunctival Squamous Cell Carcinoma (CSCC) is considered to be uncommon worldwide with a variable incidence geo-

graphically from 0.02 to 3.5 per 100,000 (Young and Foster, 1997). On the other hand increased incidence of squamous cell carcinoma of the conjunctiva with a more aggressive course is recognized among patients in tropical/thermal regions due to environmental factors (Zimmerman, 1964).

Tabbara and his group studied 10 cases of metastatic squamous cell carcinoma of the conjunctiva in Saudi Arabia. The aggressive behavior of this tumor among the Saudi population was evident by the higher number of advanced tumor and the increased frequency of metastasis and tumor-related deaths, which was attributed to the delay in seeking medical advice (Tabbara et al., 1988).

CSCC accounts for 4–29% of oculo-orbital tumors (Lee and Hirst, 1995, 1992). It has a wide spectrum of presentation, therefore all suspicious lesions should be biopsied (Meha and Fay, 2009). It is regarded as a low grade malignancy with recurrence rates that are higher for severe grades of squamous cell

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carcinoma and for cases with positive margins related to inadequate surgical excision (Tabin et al., 1997). A significantly increased risk of recurrence is also reported for older patients, large diameter and high proliferation index of the tumor (McKelvre et al., 2002).

Squamous Spindle Cell Carcinoma (SSCC) of the conjunctiva is an infrequent variant with distinct behavior which is thought to be more locally aggressive than ordinary CSCC (Cohen et al., 1980; Seregard and Kock, 1995). We are presenting a locally aggressive case of SSCC with distinct histopathological appearance and further distant metastasis.

2. The case

A 72-year-old Saudi female presented to our hospital for the first time 6 years ago with a suspicious limbal lesion in the right eye. She gave history of an excised squamous cell carcinoma at the same site 8 years before in another institution. The recurrent lesion was excised and islands of epithelial squamous cells were observed, therefore diagnosed as a completely excised recurrent limbal squamous cell carcinoma with clear surgical margins. Focal tumor areas of poorly differentiated pleomorphic, proliferating spindle-shaped cells in continuity with the overlying abnormal epithelium were overlooked by the pathologist at that time (Fig. 1). The patient was lost to follow up for 2 years, after which she presented with a painful growing limbal lesion in the same eye. Her examination revealed vision of counting fingers near the face in that eye, a vascularized nasal limbal mass extending for 3 mm over the cornea and adjacent corneal scarring (Fig. 2). Her MRI showed infiltration of this mass into the anteromedial portion of the right globe with possible penetration of the sclera (Fig. 3). The anterior exenteration specimen showed histopathologic evidence of deep corneal invasion near the limbus by poorly differentiated squamous cells (Fig. 4A and B) which stain positive with cytokeratin (Fig. 5). Three years later the patient presented to another tertiary care centre with bilateral lung masses. The biopsy of one of the lung nodules showed metastatic poorly differentiated squamous cell carcinoma with similar appearance to her original tumor (Fig. 6).

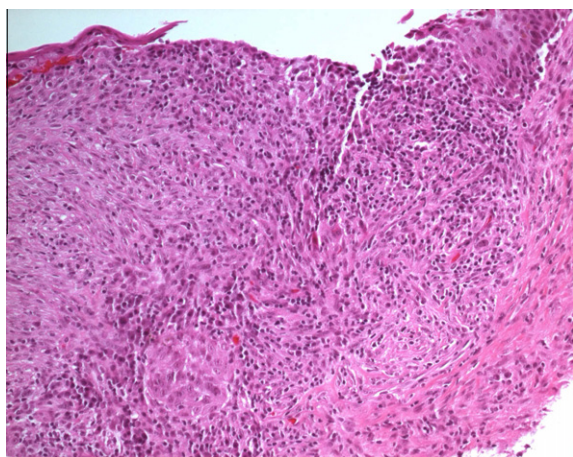


Figure 1 The histopathologic appearance of the tumor first recurrence (Hematoxylin & Eosin $\times 200$).

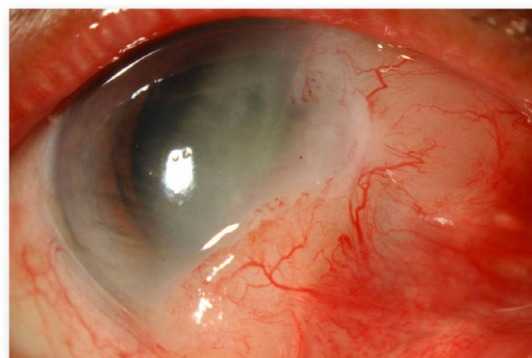


Figure 2 The vascularized recurrent mass at the limbus of the right eye.

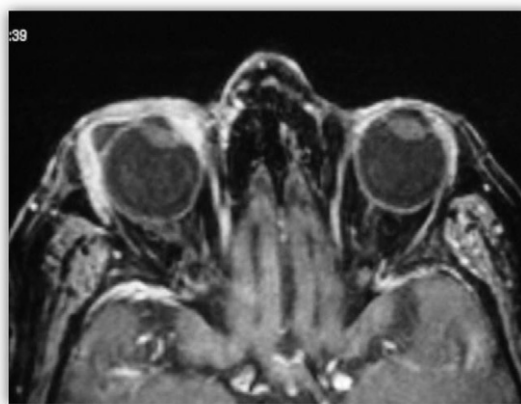


Figure 3 Magnetic resonance imaging of the right orbit showing globe invasion by the tumor.

3. Discussion

CSCC are generally non-aggressive tumors. Cases received at Armed Forces Institute of Pathology from 1928 to 1957 were followed up for 5 years or more after the initial histologic diagnosis. Their 87 cases showed evidence of regional nodal metastasis in 3 patients only who survived following local excision and/or radical neck dissection. On the other hand, local orbital recurrence with intracranial invasion has resulted in a single mortality (Zimmerman, 1964).

This behavior however is thought to be different in thermal or tropical regions such as Saudi Arabia where CSCC is expected to have an increased incidence with more advanced and aggressive cases partly due to environmental factors such as UV light exposure (Zimmerman, 1964; Tabbara et al., 1988). This was further documented in the cases studied by Tabbara and his group in 1988 including 10 patients with regional and distant metastasis. All his patients exhibited delay in management of their lesions (Tabbara et al., 1988). One of his patients developed distant metastasis in the bone and lung. However, no detailed histopathologic appearance of the original tumors was included in their study.

In regard to the histopathologic features, SSCC is a poorly differentiated variant of squamous cell carcinoma which is considered to be more aggressive and can also affect the pro-

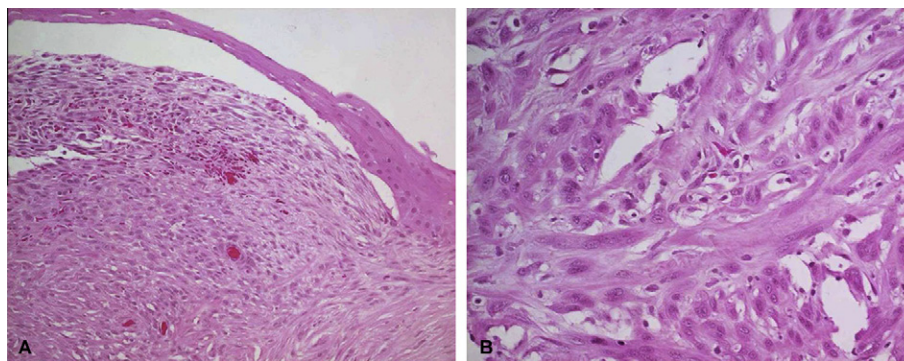


Figure 4 (A) Invasion of the corneal stroma by the recurrent tumor (Hematoxylin & Eosin $\times 200$). (B) Higher magnification of the proliferating spindle-shaped tumor cells (Hematoxylin & Eosin $\times 400$).

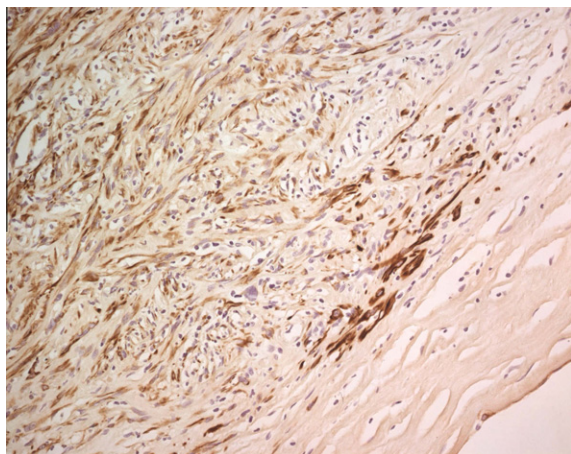


Figure 5 Positive immunohistochemical staining of the tumor cells (Cytokeratin $\times 400$).

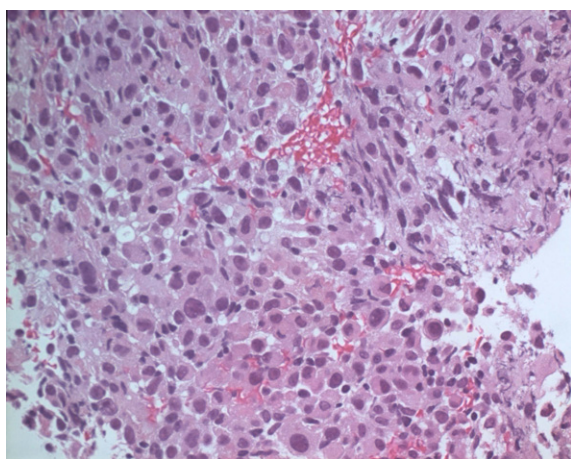


Figure 6 The histopathologic appearance of the lung metastatic poorly differentiated squamous carcinoma (Hematoxylin & Eosin $\times 400$).

gress and outcome of this disease. Seregard and his co-author presented the first case of SSCC metastatic disease or tumor-related death where the patient developed bilateral lung metastasis and rib lesions (Seregard and Kock, 1995). They agreed on the difficulty in the histologic diagnosis of this entity where

it can be confused with other lesions, and suggested that some spindle cell variants of CSCC may express cytokeratin staining, which might aid in the confirmation of the diagnosis (Seregard and Kock, 1995; Grossniklaus et al., 1987). The histopathologic slides of our patient's first recurrent local tumor were reviewed by a second pathologist to confirm the diagnosis of SSCC. This final diagnosis was not initially made until she developed the second recurrence with corneal invasion.

In spite of appropriate management of her first local recurrence then her locally invasive disease by complete surgical excision then anterior exenteration correspondingly, she eventually developed bilateral lung metastasis, became oxygen dependent and was finally kept on palliative care until she died 3 months after the diagnosis of her metastatic lung disease.

The poor prognosis and the aggressiveness in our case can be multi-factorial owing to the patient's delay in seeking medical treatment in more than one occasion along the course of her disease and that her invasive CSCC tumor proved to be of the spindle variant.

In conclusion, we recommend careful histopathologic study of excised CSCC. Identification of SSCC can help as a prognostic indicator and might necessitate closer observation and follow up of such patients. Further detailed review of the histopathological patterns of CSCC and the long-term prognosis in affected patients among the Saudi population is needed.

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