MEDICAL SCIENCE

An advanced Sebaceous Gland Carcinoma of the eyelid: A rare case report

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ABSTRACT

Sebaceous gland carcinoma (SGC) of the eyelid is an extremely lethal tumor if left untreated due to its extensive capacity to infiltrate surrounding tissue and metastasize. We hereby report a case of a 60-year-old male who presented with an ulcerative lesion over the left eye involving both eyelids and complete loss of vision for 1 year. CECT head and neck are suggestive of heterogeneously enhancing soft tissue density lesion with areas of necrosis within it seen in the left periorbital region. The lesion is causing erosive destruction of the lateral wall of the left orbit and involving the extra and intraconal compartment with ipsilateral enlarged parotid and cervical lymph nodes. Eyelid biopsy confirmed diagnosis of sebaceous carcinoma. The case was discussed in the Tumor board, and was advised upfront surgery. The patient underwent left extended orbital exenteration with left total parotidectomy, ipsilateral modified radical neck dissection and free anterolateral thigh flap reconstruction. Post-operative histopathology suggests SGC with parotid nodes and level IB, IIA lymph nodes positive for metastasis. Postoperatively patient underwent adjuvant radiotherapy. The patient is currently under regular follow-up and is disease free.

Keywords: Sebaceous gland carcinoma (SGC), Eyelid, Orbital exenteration, Modified radical neck dissection (MRND), Free anterolateral thigh flap.

1. INTRODUCTION

Sebaceous gland carcinoma (SGC) is an extremely rare and highly aggressive tumor of sebaceous glands (Kass and Hornblass, 1989). Among all malignant tumors, SGC accounts for 1-3% and approximately 0.6-10.2% of tumors of the eyelid (Yin et al., 2015). The Chinese and Japanese populations have a higher prevalence of SGC than India and Singapore; this variable prevalence among different ethnical groups explains such a large range. Recently SGC has become the most common ocular malignancy (37%) in India crossing squamous cell carcinoma (SCC) (21%) (Gupta et al., 2017). Whereas among Caucasians, ocular SGC is identified in less than 1-5.5% (Kass and Hornblass, 1989; Dasgupta et al., 2009). Early diagnosis and aggressive management are key as SGC are prone to loco-regional spread and distant metastases, with a high possibility of recurrence. According to more recent reports, the earlier

mortality rate due to SGC, which used to be around 50%, has significantly reduced to approximately 2-10% due to better knowledge about the disease (Shields et al., 2004).

2. CASE REPORT

A 60-year-old male presented with an ulcerative lesion over his left eye for one year. On thorough history taking, he had initially noticed a swelling over the left upper eyelid, which was insidious in onset and gradually progressing to involve both eyelids. The patient also complains of complete loss of vision in the left eye for the past three months patient denies any history of associated pain, trauma, redness, or itching over eyelids. The patient had no known comorbidities or previous history of periorbital irradiation. On clinical examination, an ulcero-proliferative lesion involving both the upper and lower eyelid was noted (Figure 1).

The anterior segment of the eye appeared distorted and could not be visualized. Contrast-enhanced computed tomography scan of the head and neck was suggestive of heterogeneously enhancing soft tissue density lesion with areas of necrosis within it seen in the left periorbital region. The lesion is causing erosive destruction of the lateral wall of the left orbit and involving the extra and intraconal compartments (Figure 2, 3). There are enhancing lymph nodes seen in the left preauricular region with necrosis within and bilateral submental, submandibular region along with a heterogeneously enhancing lesion in the left parotid gland with necrotic area within it suggestive of a skip lesion.

An excisional biopsy of the eyelid was done, which was suggestive of sebaceous carcinoma of the eyelid. This case was presented in the tumor board discussion and was advised upfront surgery followed by adjuvant radiotherapy. The patient underwent left extended orbital exenteration with lateral orbital wall removal, left total parotidectomy, ipsilateral modified radical neck dissection and free anterolateral thigh flap reconstruction.

An incision of full depth was made along the orbital rim, maintaining a 1cm distance from the hardened skin margin, and was carried around in a circle. The periosteum was incised all around using an electrocautery device once the orbital rim was accessed. The periosteum was then separated from the bony orbit all around using a Freer periosteal elevator. This subperiosteal dissection was extended all the way to the orbital apex after lifting the periosteum circumferentially. The supraorbital neurovascular bundle was located and then cauterized. Keeping to the medial side of the lacrimal sac, subperiosteal dissection was performed from the front to the back of the lacrimal crest. On the lateral side, after identifying the frontozygomatic suture, the zygomatico-facial and zygomatico-temporal neurovascular bundles were cauterized. The nasolacrimal duct was divided using diathermy. The blood vessels near the inferior orbital fissure were divided using electrocautery.

A pair of curved enucleation scissors were inserted into the back of the orbit once the periosteum had been detached all around, from the orbital rim to the apex. The optic nerve, superior orbital contents, and posterior orbital tissues were severed. The zygomatic bone was also removed, along with performing the orbitectomy on the superior, lateral, and inferior sides. Bone cuts were made from the superior orbital rim to the superior orbital fissure, and from the inferior orbital rim to the inferior orbital fissure at the back, and by severing the zygomatico-temporal suture. The surgical specimen is mobilized after the division of the surrounding soft tissues with electrocautery. The superficial and deep lobes of the left parotid gland were excised, preserving the facial nerve and its branches. Ipsilateral MRND was performed, and parotid nodes along with level I, II, III, and IV nodes were excised (Figure 4).

A 21 x 7 cm flap is planned over the left lateral thigh. A line marked from ASIS till the lateral border of the patella, incision marked 2 cm anterior and parallel to this line. Incision made and deepened through subcutaneous tissue and fascia incised. Rectus femoris muscle was identified and the plane between rectus and vastus lateralis entered. A single large musculocutaneous perforator was seen 16 cm from the ASIS. The perforator is dissected till the origin from the descending branch of the lateral circumflex femoral artery. The pedicle was dissected till sufficient length was achieved. A posterior incision was made, and the flap islanded, bleed checked, and was found to be bright red. The flap was detached. Flap inset into the skin defect and de-epithelized part used to fill the orbital cavity. Anastomosis of the flap artery to the left facial artery and flap vein to the left IJV tributary was done. A drain was inserted, and the neck was closed in a single layer.

The histopathological report was suggestive of moderately differentiated SGC of the eyelid, with all margins negative for malignancy and positive level IB and IIA lymph nodes (Figure 5). The postoperative period was uneventful (Figure 6). The patient was started on adjuvant radiotherapy and is currently under regular follow-up for the past six months and is disease free.

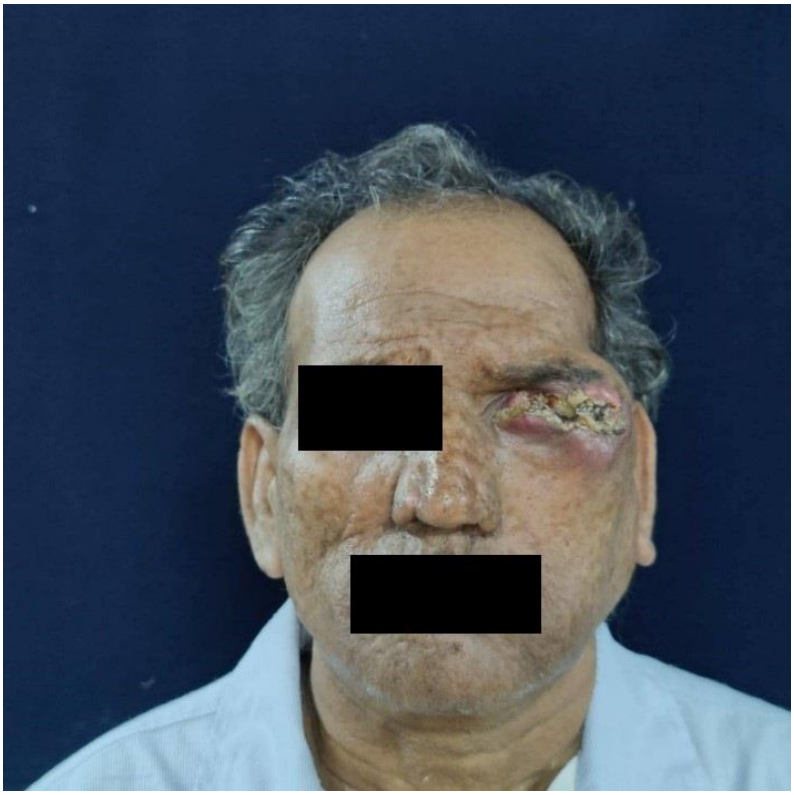

Figure 1 Preoperative clinical image showing ulcerero-proliferative lesion involving left eyelids

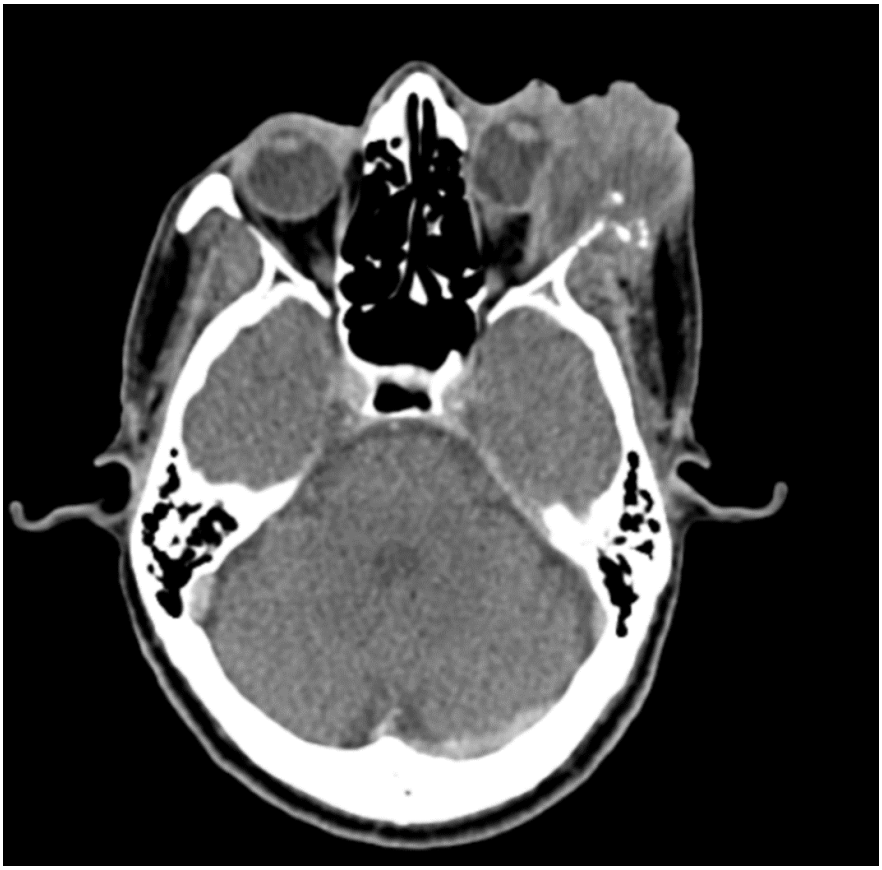

Figure 2 CT head and neck axial view showing left priorbital lesion involving intraconal and extraconal compartments

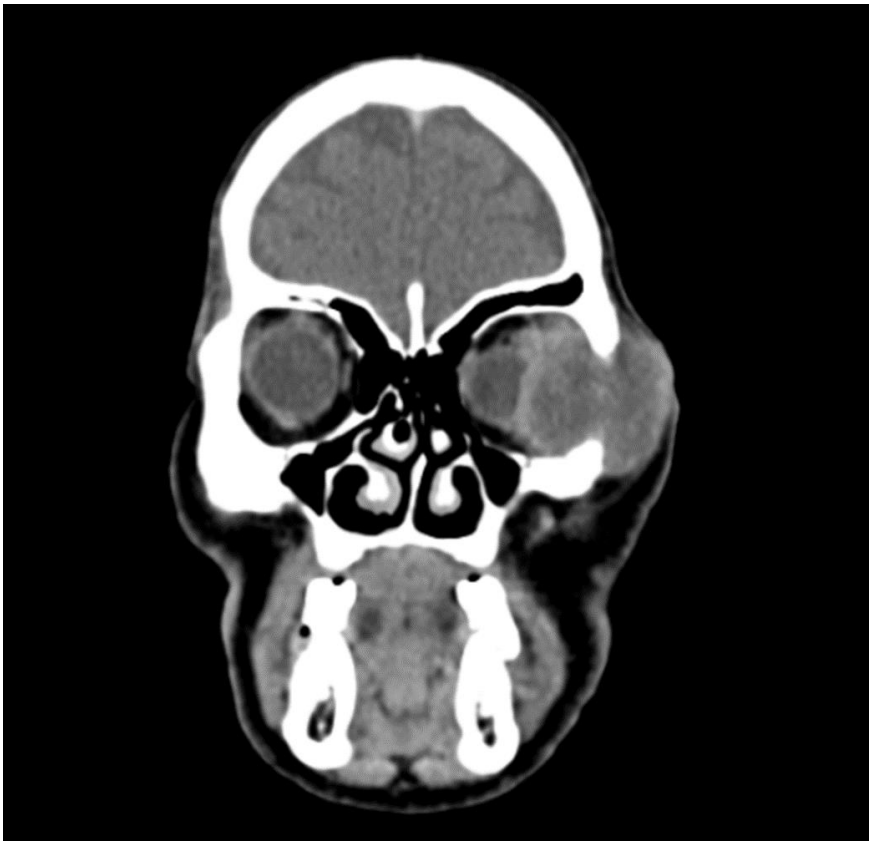

Figure 3 CT head and neck coronal view showing left periorbital lesion sowing erosion of the lateral orbital wall

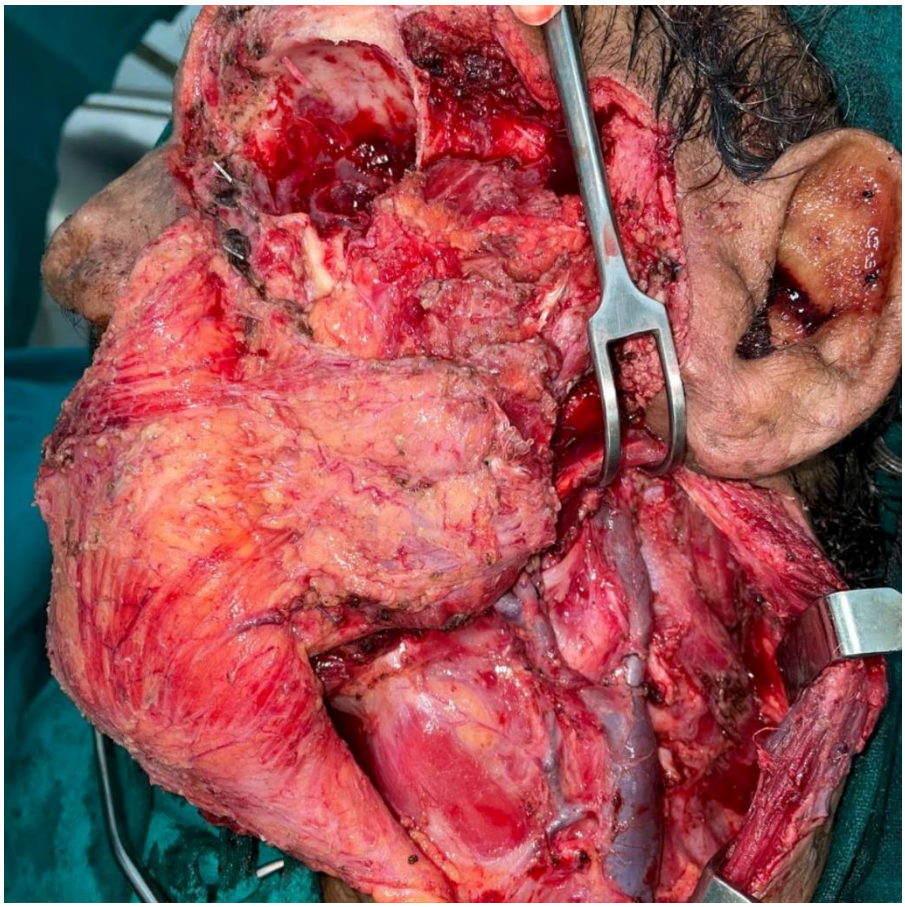

Figure 4 Intraoperative image showing left extended orbital exenteration, total parotidectomy and mrnd

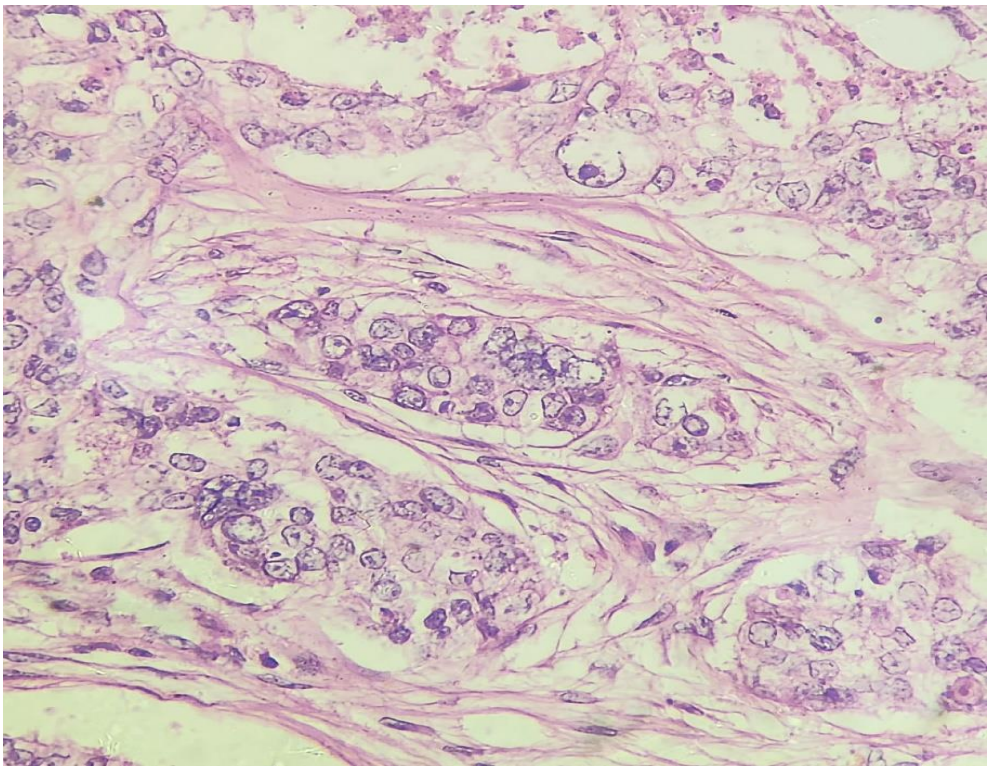

Figure 5 40x histopathological image showing sheets of pleomorphic cells with abundant foamy, finely vacuolated cytoplasm and well-defined cellular outline

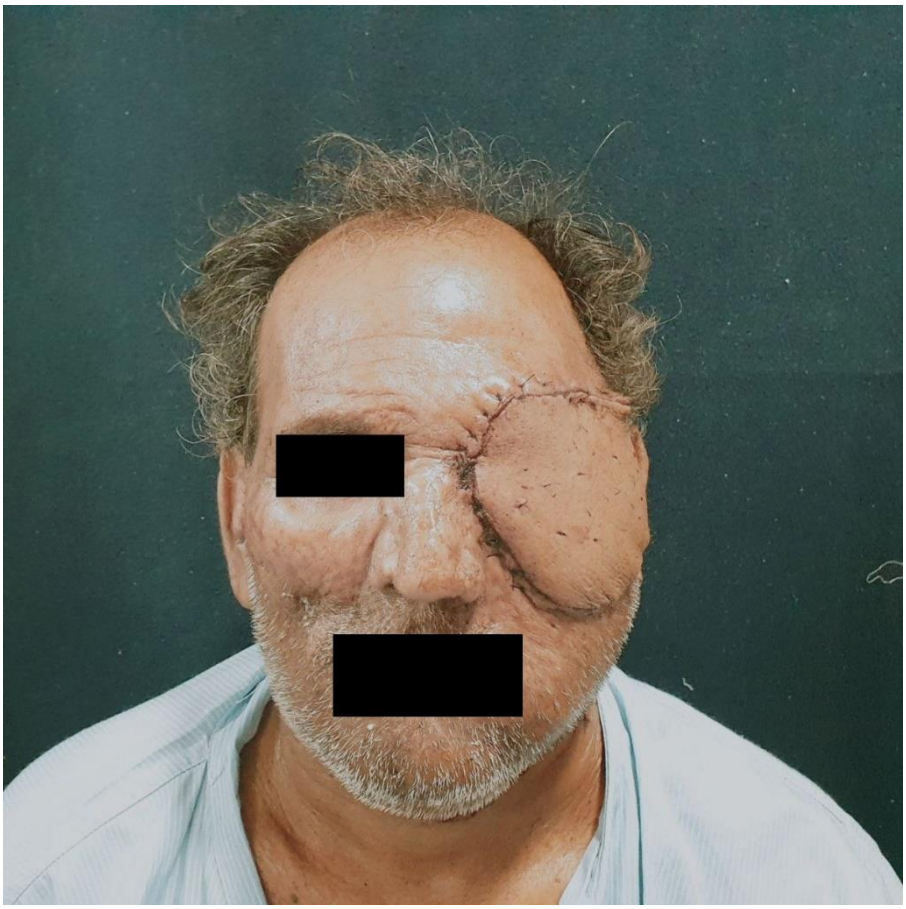

Figure 6 Postoperative clinical image showing good uptake of free anterolateral thigh flap

3. DISCUSSION

SGC classification, as per the SEER study, is ocular and extraocular type (Tryggvason et al., 2012). Extraocular sites mainly include hair-bearing areas, for example, the scalp, trunk, and genitalia, which contribute only about one-fourth of the cases of SGC (Bailet et al., 1992). Ocular SGC is mainly found to originate from the upper eyelid (39-50%) in comparison to the lower eyelid due to the Meibomian glands predominance in the upper eyelid (Dasgupta et al., 2009; Song et al., 2008). Another possible origin could be the Glands of Zeis, which are located at the eyelid margin (10%), at the caruncle (less than 10%), or in the skin of the eyebrows.

SGC commonly develops in the 6th–7th decade of life, but SGC among younger age groups has been documented (Cicinelli and Kaliki, 2019). Risk factors predisposing to the development of SGC include recurrent chalazion, history of previously taking periocular radiation, immunosuppression, Muir–Torre syndrome (MTS), familial retinoblastoma, HIV, and HPV infection (Gauthier et al., 2014). Ocular SGC can have extremely variable clinical presentation hence end number of benign and malignant conditions should be considered as differential diagnosis, including chalazion, superior limbic keratoconjunctivitis, posterior blepharitis, keratitis, SCC, basal cell carcinoma (BCC) or Merkel cell tumors (Ha and Kim, 2016).

Ocular SGC classically presents as a firm, painless, indurated thickening of the eyelid, with a yellowish hue due to the high concentration of intracellular lipids. A proliferative growth or nodular lesion could be appreciated on the eversion of the eyelid. Due to its pagetoid spread, unilateral blepharoconjunctivitis, loss of lashes, fornical shrinkage due to diffuse infiltration of the palpebral or the bulbar conjunctiva, and invasion of the cornea can be present.

The different imaging techniques currently used in the evaluation of ocular malignancies are CT and Magnetic resonance imaging (MRI), as they help in the localization of the lesion and assessing the degree of orbital involvement. MRI diffusion-weighted imaging (DWI), 18-FDG-PET CT, and MRI PET are advanced imaging modalities that are recently being used. DWI helps in differentiating benign from malignant lesions as it estimates apparent diffusion coefficient (ADC) thresholds of different lesions and staging of tumors (Sepahdari et al., 2014).

In every case of SGC, a biopsy from the involved eyelid should be taken, especially if there is a history of recurrent chalazion, madarosis, thickening of the eyelid, or ulceration over the eyelid. SGC on histology can range from well-differentiated to poorly differentiated neoplasm. Well-differentiated sebaceous carcinoma is characterized by foamy sebocytes with cytoplasmic vacuolization. Whereas cells in poorly differentiated tumors show basophilic pleomorphism and prominent nucleoli, plenty of mitotic figures, and apoptotic cells. Basaloid, epidermoid and basosquamous are the three types of cells seen in SGC (Pereira et al., 2005).

Basaloid cell type is commonly seen in infiltrative lesions, and the basaloid cell percentage is inversely proportional to the level of differentiation. SGC cytoarchitecture can be infiltrative or nodular. SGC shows “pagetoid” spread, which is the diffusion of malignant cells to the adjacent epithelium separated from the main tumor. Adipophilin (ADP), perilipin, TIP47/PP17, and nuclear factor XIIIa (AC-1A1) are frequently utilized immunohistochemical markers in the study of salivary gland carcinomas (SGC) (Muthusamy et al., 2006). Epithelial markers such as epithelial membrane antigen (EMA) and Ber-Ep4 are useful to differentiate SGC from SCC and BCC (Plaza et al., 2015).

Surgical excision and reconstruction of the eyelid remain the treatment of choice for SGC (Shields et al., 2005). Wide local excision (WLE) should be carefully evaluated by frozen section for margin involvement, but multifocal tumors can be tricky. The recommended minimum margin to be taken in WLE and MMS are 5-6mm and 2mm, respectively (Tan et al., 1991). Alternatively, taking a 3-5mm margin followed by paraffin sectioning and delayed reconstruction in centers with the non-availability of frozen sections is acceptable. Limited pagetoid spread can be managed with cryotherapy or mitomycin-C, whereas orbital exenteration is indicated for diffusely spread disease with globe and orbital involvement (Shields et al., 2015).

Achieving margin negativity should be the primary goal of orbital exenteration. In patients with metastases to regional lymph nodes, radical neck dissection followed by adjuvant radiotherapy is indicated (Husain et al., 2008). A primary treatment with radiation (50-66.6 Gy) over the tumor site or as adjuvant therapy post orbital exenteration is recommended (Hata et al., 2012). 5-fluorouracil and carboplatin/cisplatin-based neo-adjuvant chemotherapy in advanced cases of SGC may give room for possible local resection rather than more invasive procedures (Priyadarshini et al., 2010).

4. CONCLUSION

Ocular SGC, even though being a rare condition, should be considered a differential diagnosis in suspicious lesions of the eyelid as they are extremely aggressive tumors and have poor prognoses. The main objective for a surgeon should be to identify the disease correctly in its early stages and treat it prior to loco-regional spread or metastasis. It is important to raise awareness about eyelid

malignancies among the public and encourage routine eye check-ups, which can prevent loss of vision and extensive surgeries such as orbital exenteration. This ultimately improves the quality of life and morbidity related to the disease.

Author Contributions

Rahul Rajendran has collected information and prepared the manuscript, which was reviewed by all authors.

Informed consent

Not applicable.

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Conflict of interest

The authors declare that there is no conflict of interest.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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