Case Report

Orbital sebaceous gland carcinoma: A brief communication

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ABSTRACT

Sebaceous gland carcinoma commonly arises in the periocular area and is an uncommon condition. Its orbital origin is even rare with isolated reports in literature search. Its early diagnosis is frequently missed owing to the subtle presentation that mimics various benign conditions. Surgery with wide resection margin is considered the standard of care. Irradiation is frequently indicated and administered as an adjuvant regimen following surgical resection. The role of chemotherapy in this disease remains investigational and is usually employed in recurrent settings.

Key words: Masquerade syndrome, Orbital sebaceous gland carcinoma, radiotherapy

INTRODUCTION

Sebaceous gland carcinoma (SGC) is a rare, highly malignant and potentially lethal tumor that arises from meibomian glands of the tarsal plate, glands of Zeis or from sebaceous glands of the caruncle, eye brow or facial skin.[1] SGC of the eyelid is known for masquerading as a benign or malignant condition ("masquerade syndrome") often causing a delay before the correct diagnosis. The misleading clinical manifestations may suggest inflammation, including unilateral conjunctivitis, blepharitis, tarsitis, blepharocojunctivitis, and keratoconjunctivitis.[2] It may also mimic a sebaceous adenoma and is included in the group of simulating lid lesions (inclusion cyst, papilloma, senile keratosis, keratoacanthoma, benign keratosis, dermoid cyst, and amyloidosis).[3] SGC arising in the orbit is an extremely rare happenstance and has been reported sporadically. These tumors are reported to behave aggressively with the tendency to metastasize early with higher mortality rates especially if there is a delay in the diagnosis.[4] Data in the literature indicate that it has a 30–40% risk for recurrence, 20–25% for metastases and 20% for tumor-related mortality.[2]

Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.148981

CASE REPORT

A middle aged male in his sixth decade, with no known co-morbidities presented to us with the chief, complains of diminished vision in right eye, restriction of ocular movements and occasional diplopia. He was in a good general condition with an eastern cooperative oncology group performance status of 1 at presentation.

The patient had a 2 months history of blurring of vision associated with redness of the right eye. Earlier clinical examination had shown right eye proptosis (upward and outward) along with a large firm to hard mass palpable inferior and lateral to the globe. Fundus disks were normal. He was further evaluated with a contrast enhanced computerized tomography (CT) scan [Figures 1 and 2] that revealed a well-defined soft tissue extraconal mass lesion measuring 25 mm × 30 mm × 21 mm at the infero-medial aspect of right orbit. Following this he underwent an anterior orbitotomy and mass debulking.

The patient presented to our institute, and a histopathological review revealed sebaceous carcinoma with fibroadipose tissue infiltration along with brisk mitosis. Resection plane was found to be involved with the disease. He was further subjected to a magnetic resonance imaging of the eye that revealed soft tissue edema with small residual enhancing

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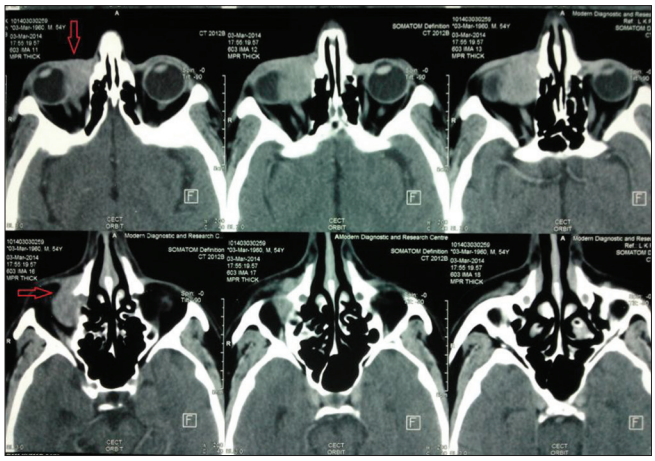

Figure 1: Preoperative computerized tomography (axial) showing a soft tissue extraconal mass lesion at the infero-medial aspect of the right orbit

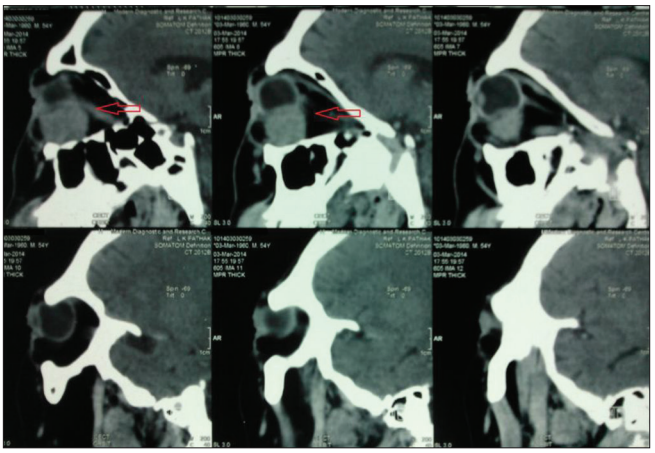

Figure 2: Preoperative computerized tomography (sagittal)

lesion along the infero-medial aspect of the right orbit with contiguous enhancement of the right inferior and middle rectus muscles. A whole body 18 fluorodeoxyglucose (FDG) positron emission tomography CT was done to rule out any distant metastasis and revealed no evidence of FDG avid regional lymph nodes (LNs) or any distant metastases.

The patient underwent a revision surgery in the form of a right radical orbital exantration with extended neck dissection with temporal reconstruction. Surgical pathology revealed poorly differentiated sebaceous carcinoma involving orbital soft tissues [Figures 3 and 4]. Lymphovascular emboli and perineural invasion were noted. The eyeball, skin, and optic nerve were free of the tumor. All (deep/rectus muscle) margins were free of tumor deposits except the medial margin. Parotid gland parenchyma was free. One intraparotid LN out of four showed metastases. None of the 22 resected neck nodes showed metastases.

In accordance with the standard protocol, he was then referred to us for adjuvant radiotherapy (RT). At presentation, he had recovered well from his surgery with mild difficulty in opening mouth along with mild-moderate pain at the operated sites. The surgical wound had healed well. Patient was planned to receive a total dose of 6000 cGy to be delivered in 30 sessions to the clinical target volume 1 (CTV1) that comprised of the primary tumor bed and received a simultaneous dose of 5500 cGy in 30 sessions by Simultaneous Integrated Boost technique to the CTV2 that included CTV1 along with nodal drainage in right temporal region, right parotid and right level II cervical region. A final safe margin was added to the CTV to form the planning treatment volume to take care of microscopic disease as well as any inadvertent set-up variations. He was treated with intensity modulated RT technique using five beams and 32 segments [Figures 5 and 6]. He tolerated the treatment well and completed the prescribed course of RT without any treatment gaps.

After completion of his adjuvant RT, he presented for follow-up after 3 months and was found to have no evidence of any residual/recurrent disease.

DISCUSSION

Sebaceous gland carcinoma is a rare malignant tumor originating from adnexal skin structures. Most of these malignancies develop in the head and neck area, notably in the eyelid. Other sites include the parotid and submandibular glands, the external auditory canal, the trunk or the limbs.[5] Orbital SGC is considered to be potentially aggressive, with local recurrence rates ranging from 11% to 23%. Regional and distant metastases have been reported in up to 21% of cases. Tumors of this type commonly reveal a diffuse and invasive growth pattern and are associated with a high incidence of loco regional recurrence in the skin and LN but also with distant metastases. Search in PubMed reveals isolated case reports of orbital SGC depicting its rare and sporadic presentation.[6-8] Surgical excisions and LN dissections remain the standard of care whenever involvement is suspected. Retrospective studies have reported an overall 5 years survival of approximately 70% regardless of where the primary tumors had been located.[9] Regional lymphadenectomy plays an important role in the management of SGC since LN involvement is seen in approximately 15–21% of cases.[10] RT is more commonly used as an adjuvant than a primary treatment. In ocular SGC, postsurgical local irradiation has been shown to improve local control and to result in satisfactory cosmetic outcomes.[11]

Patients with SGC must be followed-up at short intervals postoperatively as the tumor has a fast growth potential. Adequate follow-up includes meticulous inspection of the local site. Palpation of the preauricular, submandibular and other neck LN chains is mandatory. The approximate guidelines are 3 monthly interval during the 1st year,

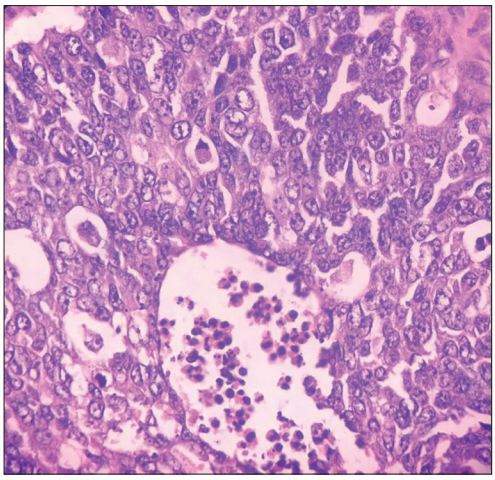

Figure 3: Section shows irregular lobules and sheets of malignant cells (H and E, ×100)

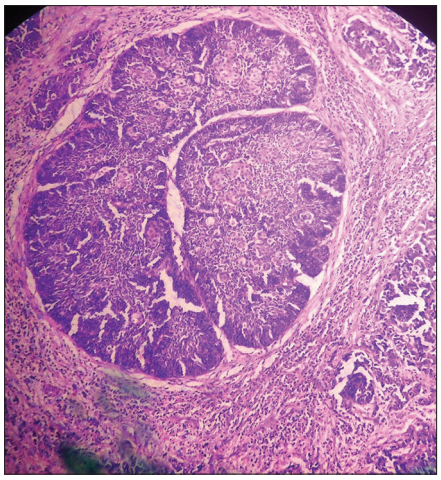

Figure 4: Section shows undifferentiated atypical cells with considerable nuclear and nucleolar pleomorphism with eosinophilic cytoplasm and mitotic figures (H and E, ×400)

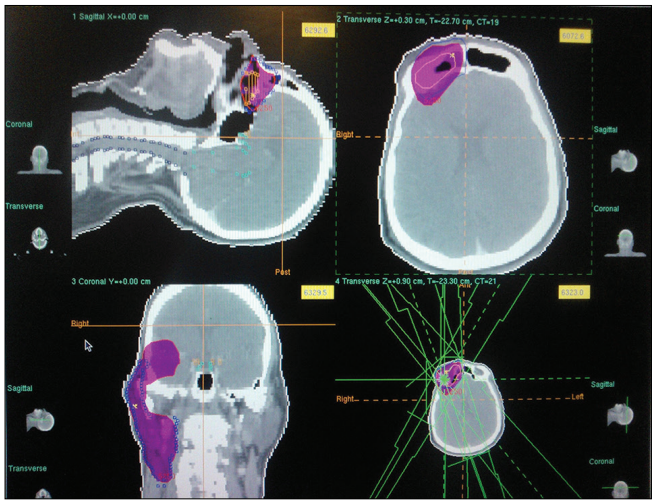

Figure 5: Radiotherapy plan image

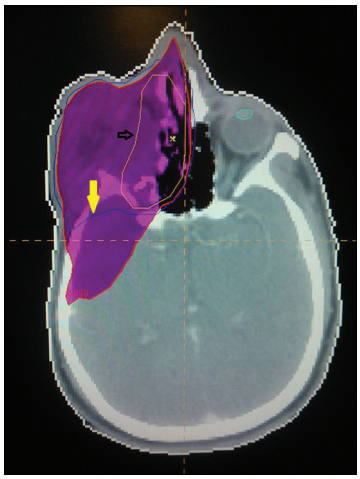

Figure 6: Radiotherapy dose distribution. Black arrow showing coverage of the primary tumor bed to a dose of 6000 cGy clinical target volume 1 (CTV1). Yellow arrow showing CTV2 coverage to a dose of 5500 cGy

6 monthly during the 2nd year and then on a yearly basis for life.[12]

CONCLUSION

Sebaceous gland carcinoma is a rare tumor of the eyelid that can mimic many inflammatory lesions. Its orbital origin makes the presentation even rarer. Imaging modalities are mandatory to assess the local involvement as well as to rule out distant metastases thereby guiding the appropriate modality of treatment. Surgery is the standard of care while irradiation is frequently administered as an adjuvant regimen following surgical resection.

ACKNOWLEDGMENT

Dr. Shalini Goel, Attending Consultant, Department of Pathology, Medanta - The Medicity.

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Cite this article as: Tiwari V, Pande SC, Verma K, Goel S. Orbital sebaceous gland carcinoma: A brief communication. Clin Cancer Investig J 2015;4:465-8.

Source of Support: Nil, **Conflict of Interest:** None declared.