Secondary mucoepidermoid carcinoma of the orbit

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A 40-year-old man presented with right eye axial proptosis and ophthalmoplegia for 3 months. Imaging study showed a right intraconal mass with the erosion of the orbital floor. Incisional biopsy revealed mucoepidermoid carcinoma. Nasal endoscopy was normal and systemic tumor screening was negative for a primary source. The patient underwent right orbital exenteration, uncinectomy, nasal and maxillary mucosal biopsy. Malignant cells were found present in the mucosa of maxillary sinus roof and uncinate bone. The postoperative positron emission tomography scan showed residual active lesion in right orbital apex and maxilla but no primary lesion elsewhere. The patient subsequently underwent 35 cycles of postoperative radiotherapy. Primary mucoepidermoid carcinoma of the orbit is rare and typically arises from the lacrimal gland or sac. Those tumors not arising from lacrimal apparatus should be presumed metastatic in origin, and the thorough systemic survey should be undertaken in the search for the primary tumor.

Key words: Metastatic, mucoepidermoid carcinoma, orbit

Metastasis to orbit accounts for 1–13% of all the orbital tumors.[1] Its occurrence in patients with known systemic carcinoma is approximately 2–3%.[1] The primary tumor sites include breast, prostate gland, lung, skin, kidney, gastrointestinal, choroid, parotid, and adrenal gland. In 10% of the patients, the primary tumor remained undetected despite extensive systemic evaluation. The prognosis is generally poor.[1]

We report a rare case of secondary high-grade invasive mucoepidermoid carcinoma in the orbit. The probable primary tumor site was presumed to be maxillary sinus.

Case Report

A 40-year-old man presented with right eye swelling for 3 months duration. It was associated with diplopia and headache. Patient's vision was 20/30 bilaterally with grade one relative afferent pupillary defect over the right eye. Physical examination revealed right eye axial proptosis and ophthalmoplegia. Fundus examination showed a mass effect pushing forward at the posterior pole.

Imaging study showed a right intraconal mass with the erosion of the orbital floor. A mass was seen in the right retro-orbital space extending to the orbital apex [Figs. 1-3] and measured 2.1 cm × 1.9 cm × 1.9 cm. The medial and lateral rectus muscles were involved. Thickened mucosa was seen within both maxillary sinus but no mass seen.

Incisional biopsy result showed high-grade invasive carcinoma with features suggestive of mucoepidermoid carcinoma [Figs. 4 and 5]. Microscopically, there were islands and small nests with <20% intracystic component. The tumor cells were mainly epidermoid cells. They showed markedly pleomorphic vesicular to hyperchromatic nuclei. Occasional mucous cells were also present. The mucous cells stained positive with periodic acid-Schiff. Mitotic activity was 3/10 high power field (Olympus BX41). There was no necrosis noted. Perineural and lymphovascular areas were clear. The tumor was positive for p63, cytokeratin (CK) 5/6, epithelial membrane antigen, Bcl 2. Focal CK7 positive was seen within the mucous cells.

Nasal endoscopy was normal and systemic tumor screening was unremarkable for a primary source. The patient underwent right orbital exenteration, uncinectomy, nasal mucosal, and maxillary mucosal biopsy. Intraoperatively, the tumor was firmly adherent to the orbital floor and extending posteriorly to the orbital apex. There was no macroscopic tumor mass seen within the maxillary sinus. Histopathologic results were consistent with high-grade mucoepidermoid carcinoma; malignant cells were also present in the mucosa of maxillary sinus roof and uncinate bone. The tumor mass was mainly present over the posterior part of the orbit and involved extraocular muscles and bones. The globe, eyelids, and optic nerve were free of tumor cells [Fig. 6]. The lacrimal gland, canaliculi, and sac were also unremarkable. There was no lymphovascular involvement. Most of the resected margins were invaded by tumor [Fig. 7]. Armed Forces Institue of Pathology (AFIP) point system showed high-grade tumor with a score of 8 points, the presence of intracystic component <20% (2 points), necrosis (2 points), anaplasia (4 points) [Table 1].[2] Under Brandwein point system, the tumor mass scored 12 points which indicated a high-grade tumor as well. A total of 12 points were evidenced by the presence of intracystic component <25% (2 points), tumor invades in small nest and island (2 points), pronounced nuclear atypia (2 points), bony invasion (3 points), and necrosis (3 points) [Table 2].[2]

The postoperative positron emission tomography scan showed residual active lesion in right orbit and maxilla,

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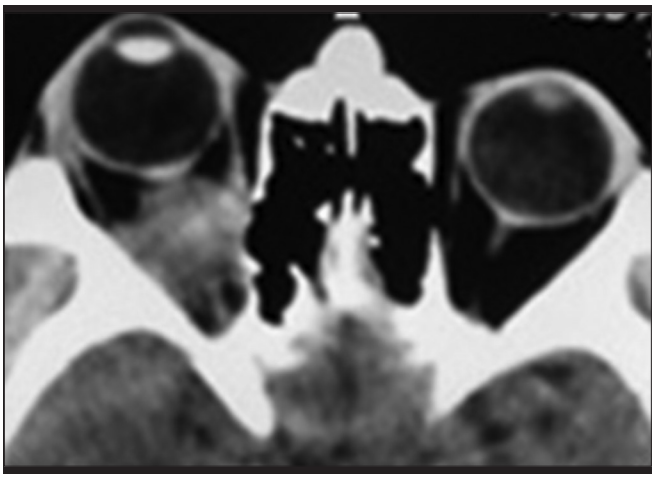

Figure 1: Plain computerized tomography scan of right orbital axial view showed a homogenous enhancing retrobulbar mass involving extraocular muscles and causing proptosis

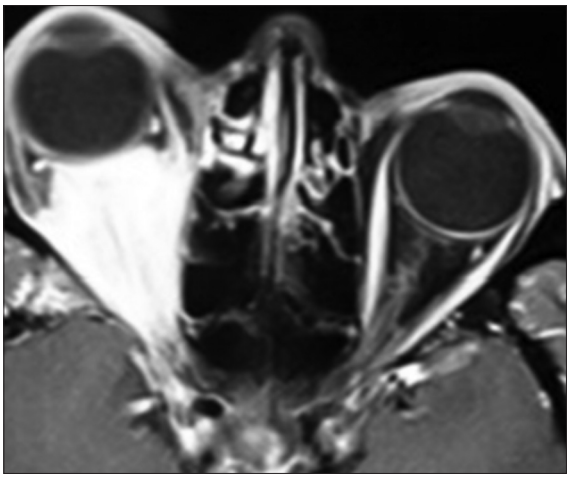

Figure 2: Contrast-enhanced T1-weighted magnetic resonance imaging of axial view of the right orbital retrobulbar mass

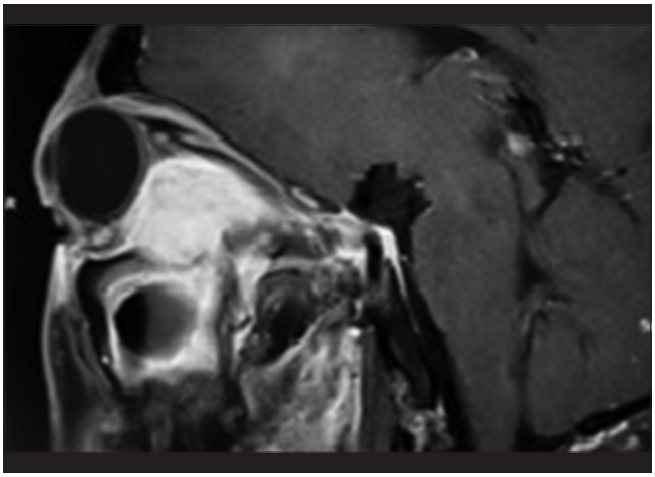

Figure 3: Contrast-enhanced T1-weighted magnetic resonance imaging of sagittal view showed right retrobulbar mass extended to orbital apex with erosion of the orbital floor

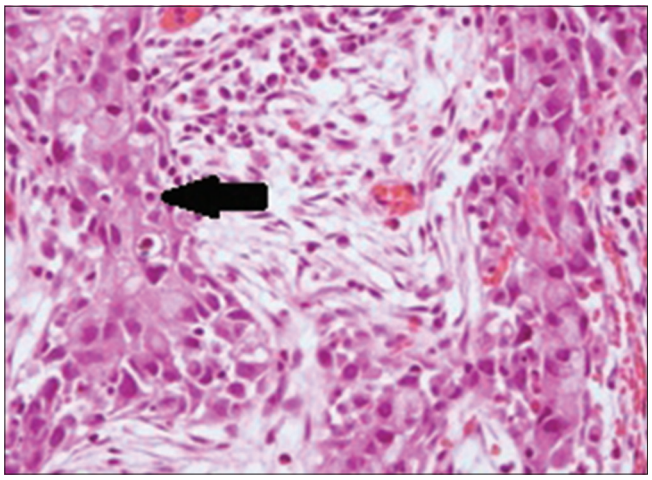

Figure 4: Histopathological examination (H and E, ×40) showed presence of mucus tumor cells (black arrow)

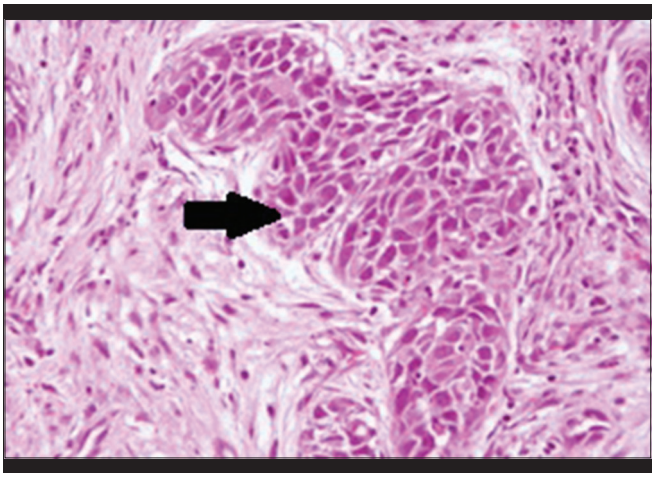

Figure 5: Histopathological examination (H and E, ×10) showed presence of epidermoid tumor cells (black arrow)

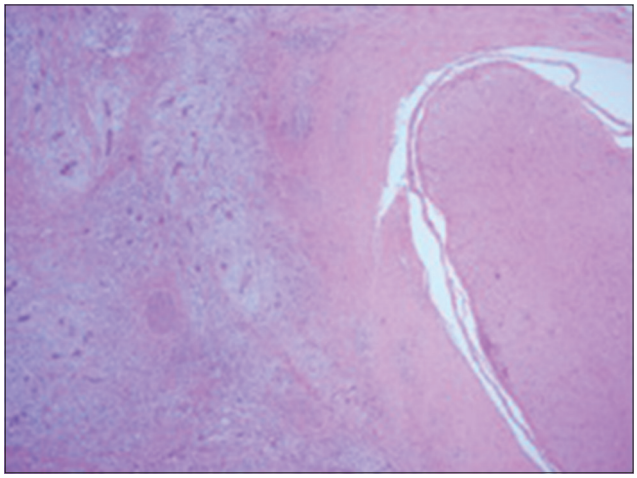

Figure 6: Histopathological examination (H and E, ×4) showed clusters of tumor cells surrounded by desmoplastic stroma (left) and adjacent optic nerve (right). The optic nerve is free from tumor

but there was no primary lesion elsewhere. The patient subsequently underwent 35 cycles of postoperative radiotherapy (70 gray). At 9 months of follow-up postradiotherapy, the patient was well and healthy and presently awaiting orbital prosthesis fitting.

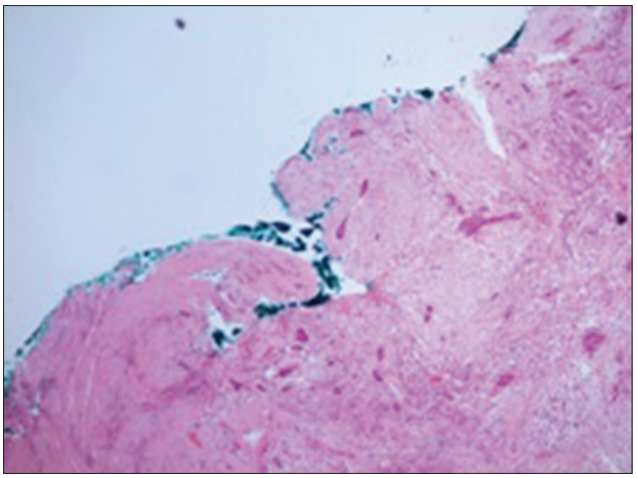

Figure 7: Histopathological examination (H and E, ×4) showed tumor cells on resection margin

Table 1: Armed Forces Institute of Pathology grading system for mucoepidermoid carcinoma

Histologic features	Point value
Intracystic component <20%	2
Neural invasion	2
Necrosis	3
Four or more mitotic figures/10 HPF	3
Anaplasia	4
Grade	**Total score (point)**
Low	0-4
Intermediate	5-6
High	7-14

HPF: High power field

Table 2: Brandwein grading system for mucoepidermoid carcinoma

Histologic features	Point value
Intracystic component <25%	2
Tumor invasion in form of small nests and islands	2
Pronounced nuclear atypia	2
Lymphovascular invasion	3
Invasion of bone	3
Perineural spread	3
Necrosis	3
Grade	**Total score**
I	0
II	2-3
III	4 or more

Discussion

Orbital malignancy can be a diagnostic challenge to the ophthalmologist. Prompt diagnosis and management is important to preserve the remaining vision and hopefully improve the survival rate.

Mucoepidermoid carcinoma is a rare tumor and commonly arises from the salivary glands in the head and neck region. Ocular and ocular adnexa involvement is rare, and the primary site of origin is usually from the conjunctiva, lacrimal gland, or lacrimal sac.[3] A rare case of mucoepidermoid carcinoma arising from conjunctiva with lung metastasis has been reported by Rishi *et al*.[4] Primary orbital mucoepidermoid carcinoma not arising from the lacrimal apparatus or conjunctiva has never been reported to our knowledge and should be presumed metastatic in origin.

It is not rare to see orbital metastasis preceding the diagnosis of a primary tumor; in fact, 19–25% of orbital metastatic cases did not have any known history of systemic malignancy. Orbital metastases typically cause mass effect and bony involvement. Extraocular movement restriction may be out of proportion to the proptosis.[5]

In this reported case, the diagnostic dilemma was that the largest and only tumor bulk was found in the orbit and hence the hunt for the primary source. In 3–5% of all tumor distant metastases, it arises at an early stage of tumor progression. Therefore, such unexpected phenotype can overcome the primary lesion's growth.[6]

There are numerous features supporting the diagnosis of a metastatic origin in this case apart from the cellular origin of the tumor. Bony involvement is typical in orbital metastasis, as evidenced by orbital floor involvement in this patient while a primary lesion of mucoepidermoid carcinoma in orbit rarely involved bone.[5,7] The tumor cells were also present in the maxillary and uncinate biopsies. This high-grade invasive carcinoma most likely originated from the maxillary sinus which had invaded the orbital floor and orbital soft tissue. Wolfish *et al*. reported a case series of 19 cases of a sinonasal tract of mucoepidermoid. Majority of the patients presented with a mass within the nasal cavity or paranasal sinus. Ophthalmologic features included diplopia, proptosis, and visual field changes. Maxillary sinus is closely in contact with the orbit and tumors arising from it may invade the orbit.[8]

Exenteration of the orbit was needed to eradicate the high grade and invasive carcinoma. This patient was at high risk of recurrence and poor prognosis in view of high grading in both AFIP and Brandwein point system, hence, postoperative radiation was indicated. The patient had undergone a total of 70 gray, 35 cycles radiation therapy under the care of an oncologist.

Treatment of orbital metastasis is always palliative. Radiotherapy is the mainstay for orbital metastases.[5]

Prognosis for orbital metastases is generally poor even if the local orbital tumor can be controlled.[9] Two years survival rate is 27%.[10] The most significant prognostic factors were the patient's age as well as a treatment method. Those aged 56 and above have a poorer prognosis. Patients who are treated with surgery with radiation had better prognosis.[11] Thus, adjunctive radiotherapy should be considered for cases with high risk of recurrence and poor prognosis.

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Conflicts of interest

There are no conflicts of interest.

References

1. Shields JA, Shields CL, Brotman HK, Carvalho C, Perez N, Eagle RC Jr. Cancer metastatic to the orbit: The 2000 Robert M. Curts Lecture. Ophthal Plast Reconstr Surg 2001;17:346-54.
2. Gale N, Zidar N. Tumors of head and neck. In: Damjanov I, Fan F, editors. Cancer Grading Manual. 2nd ed. Berlin, Heidelberg, New York: Springer; 2013. p. 24-6.
3. Dithmar S, Wojno TH, Washington C, Grossniklaus HE. Mucoepidermoid carcinoma of an accessory lacrimal gland with orbital invasion. Ophthal Plast Reconstr Surg 2000;16:162-6.
4. Rishi P, Sharma R, Subramanian K, Subramaniam N. Mucoepidermoid carcinoma of the conjunctiva with lung metastasis. Indian J Ophthalmol 2015;63:457-9.
5. Ahmad SM, Esmaeli B. Metastatic tumors of the orbit and ocular adnexa. Curr Opin Ophthalmol 2007;18:405-13.
6. Stella GM, Senetta R, Cassenti A, Ronco M, Cassoni P. Cancers of unknown primary origin: Current perspectives and future therapeutic strategies. J Transl Med 2012;10:12.
7. Schwarcz RM, Coupland SE, Finger PT. Cancer of the orbit and adnexa. Am J Clin Oncol 2013;36:197-205.
8. Wolfish EB, Nelson BL, Thompson LD. Sinonasal tract mucoepidermoid carcinoma: A clinicopathologic and immunophenotypic study of 19 cases combined with a comprehensive review of the literature. Head Neck Pathol 2012;6:191-207.
9. Hassler W, Unsold R, Schick U. Orbital tumors: Diagnosis and surgical treatment. Dtsch Arztebl 2007;104:A496-501.
10. Char DH, Miller T, Kroll S. Orbital metastases: Diagnosis and course. Br J Ophthalmol 1997;81:386-90.
11. Ozawa H, Tomita T, Sakamoto K, Tagawa T, Fujii R, Kanzaki S, *et al.* Mucoepidermoid carcinoma of the head and neck: Clinical analysis of 43 patients. Jpn J Clin Oncol 2008;38:414-8.