

Nasal epithelial myoepithelial carcinoma: An unusual cause of epiphora, a case report and review of the literature

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ABSTRACT

Introduction: Epithelial myoepithelial carcinoma (EMC) of the nasal cavity is a rare tumor, and here we describe the first case of EMC of the nasal cavity presenting with epiphora. A case presentation and review of the literature is provided.

Methods: A case report is described of a 63-year-old man who presented with unilateral epiphora and was found via a thorough history and physical examination to have a nasal tumor. The physical examination consisted of an ocular examination, including probing and irrigation, and a detailed nasal examination (anterior rhinoscopy, nasal endoscopy). The nasal examination was prompted by the patient's report of concurrent nasal symptoms during history taking. Immunohistochemistry subsequently identified the nasal tumor as EMC. A literature search was performed to gain insights into similar malignancies of the nasal cavity.

Results: Eight cases of EMC of the nasal cavity were identified in the literature, none of the patients presented with epiphora. The case presented here resulted in resolution of the patient's symptoms and no evidence of disease after surgical excision.

Conclusion: Epithelial myoepithelial is a rare salivary gland malignancy that can arise in the nasal cavity. Unilateral epiphora with concurrent nasal symptoms should prompt nasal cavity examination for the possibility of an obstructive tumor.

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Epiphora results from an imbalance between tear production and drainage.¹ Epiphora is four times more common in women than in men,² and its prevalence ranges from 9% to 10% at 50 years old to 35–40% at 90 years old.³ The differential diagnosis for epiphora is broad and includes reflex tearing from dry eye, with compensatory hypersecretion,⁴ poor tear distribution due to altered blink dynamics and lid malposition,⁵ and defective tear drainage from stenosis of the nasolacrimal drainage system.⁶ The etiology of nasolacrimal duct obstruction may be primary (idiopathic)⁷ or secondary, due to a variety of causes, including local inflammation or fibrosis (e.g., iodine-131 therapy,⁸ glaucoma drops,⁹ rhinosinusitis¹⁰), systemic inflammatory disease (e.g., Wegener granulomatosis,¹¹ sarcoidosis¹²), facial trauma,¹³ previous sinonasal surgery,^{14–18} or nasolacrimal system^{19,20} or sinonasal²⁰ neoplasia.

Sinonasal neoplasms comprise <1% of all malignancies and <3% of all malignant tumors of the upper

aerodigestive tract,²¹ and they have been estimated to occur at an incidence of 0.556 cases per 100,000 population per year.²² Epithelial myoepithelial carcinoma (EMC) is a rare type of tumor that predominantly occurs in the salivary glands²³ but also occurs rarely in the nasal cavity. Previous cases of EMC of the nasal cavity have been described as presenting only with nasal symptoms.^{24–31} Herein, we reviewed the previously reported cases of EMC of the nasal cavity and present, to our knowledge, the first reported case of a patient with EMC presenting as unilateral epiphora.

CASE REPORT

A 63-year-old man was referred to our department of otolaryngology with a 2-year history of epiphora and “sandy” feeling in the left eye. He reported a 1-year history of intermittent nasal crusting and bleeding that had become persistent. On ophthalmology examination, the left puncta appeared patent but small compared with the right puncta. Lower lid ectropion and blepharitis were noted bilaterally. Probing and irrigation demonstrated a 100% blockage of the left nasolacrimal system, with normal canaliculi, which indicated a distal obstruction. On nasal endoscopy, the nasal cavities were patent, but a friable mass was noted in the left inferior meatus (Fig. 1).

Computed tomography of the paranasal sinuses showed a 1.5 × 1.1 × 1.6 cm (anteroposterior × transverse × cranial caudal) anterior soft-tissue mass in the left nasal cavity located between the inferior turbinate and the medial wall of the maxillary sinus abutting the

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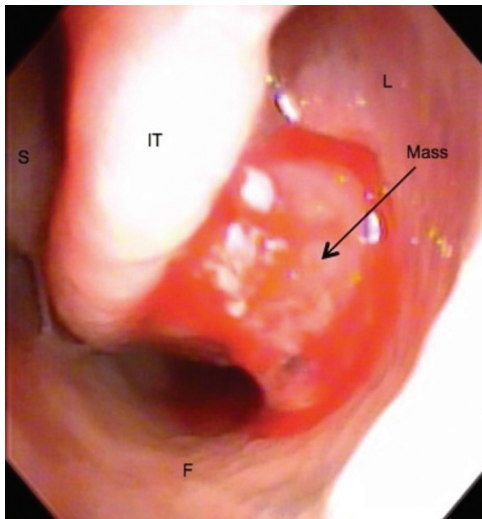


Figure 1. Mass noted in the left inferior meatus during nasal endoscopic examination. S = nasal septum; IT = inferior nasal turbinate; L = lateral wall of the nasal cavity; F = floor of nasal cavity.

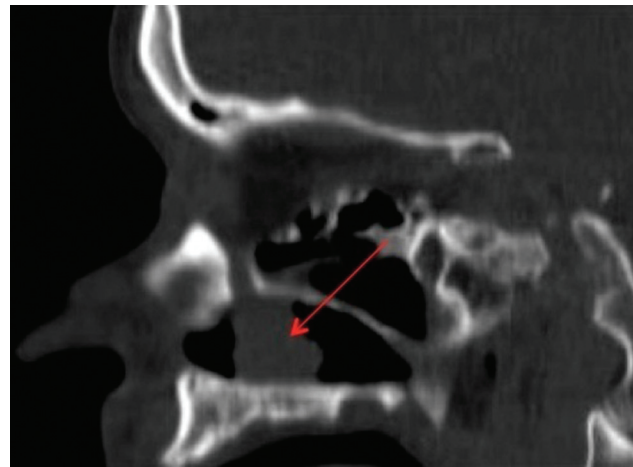


Figure 3. Sagittal computer tomography of the paranasal sinuses, showing a tumor abutting the nasolacrimal duct.



Figure 2. Axial computed tomography of the paranasal sinuses, showing a tumor abutting the nasolacrimal duct. S = nasal septum; IT = inferior nasal turbinate; M = maxillary sinus.

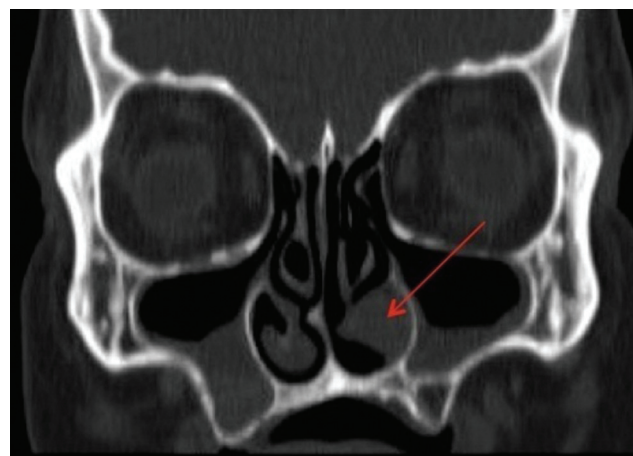


Figure 4. Coronal computed tomography of the paranasal sinuses, showing a tumor underneath the inferior turbinate.

orifice of the nasolacrimal duct. The mass was associated with medial displacement of the inferior turbinate and slight underlying bony thinning of the medial wall of the maxillary sinus (Figs. 2–4).

Endoscopic excisional biopsy of the primary mass was performed with the patient under general anesthesia. On gross examination, the primary tumor consisted of tan hemorrhagic tissue. Hematoxylin and eosin staining showed an uncapsulated epithelial neoplastic proliferation that infiltrated the submucosa. The mass consisted of a tubular glandular growth composed of a dual population of cells: inner epithelial-

appearing cells and outer myoepithelial-appearing cells with a clear cytoplasm. Oncocytic cytoplasmic changes were observed in some of the epithelial-appearing cells and clusters of predominantly myoepithelial cells without tubular glandular structures were noted. There was infiltration of the tumor into the submucosa without perineural or lymphovascular invasion. Immunohistochemistry definitively confirmed the identity of the epithelial- and myoepithelial-appearing cells. Both cell populations were positive for cytokeratin 5/6 (Fig. 5) and cytokeratin 903, and variably positive for S-100. The epithelial cells were identified by dedicated CAM 5.2 staining, whereas the myoepithelial cells were identified by dedicated α smooth muscle actin (Fig. 6), p63, and calponin staining. Ki67 staining showed a proliferation rate of 5–10%. Overall, these findings were consistent with an epithelial-myoepithelial carcinoma.

The patient underwent an endoscopic medial maxillectomy to obtain definitive margins. In addition, a

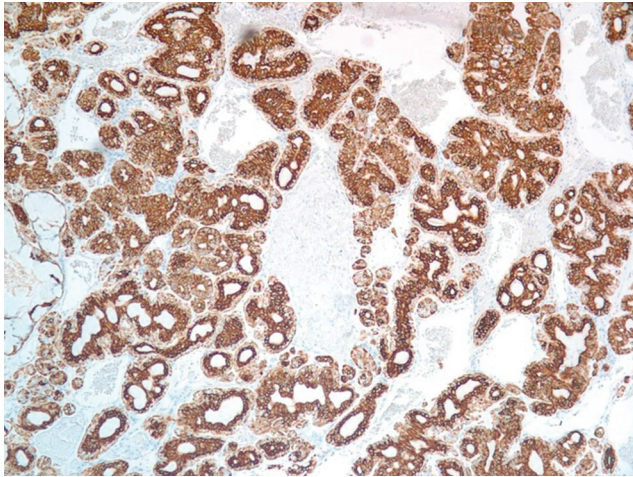


Figure 5. Inner luminal cells and outer myoepithelial cells positive for cytokeratin 5/6 (magnification $\times 100$).

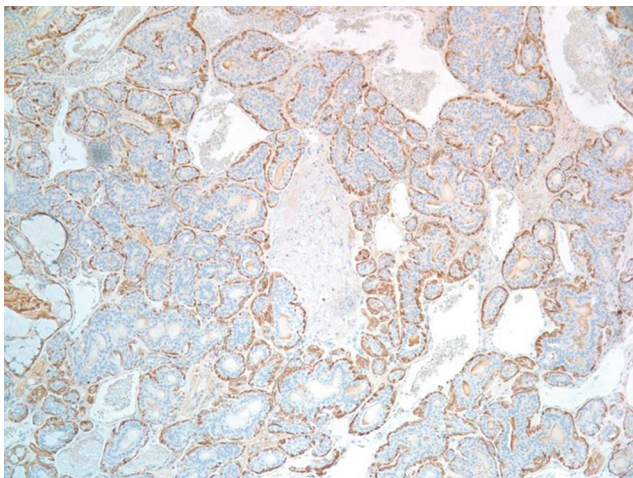


Figure 6. Outer myoepithelial cells stain positive for α smooth muscle actin (magnification $\times 100$).

prophylactic endoscopic dacryocystorhinostomy was performed to prevent potential stenosis of the nasolacrimal duct given its proximity to the initial tumor. The patient's postoperative course was unremarkable, and final pathology demonstrated no further evidence of tumor. One year after the resection, the patient was doing well, without evidence of recurrent or metastatic disease. Given the malignant nature of the disease, the patient will continue to be regularly followed up until 5 years after the resection.

DISCUSSION

History and physical examination are the keys to determine the underlying cause of epiphora.^{32–34} When evaluating a patient with epiphora, physicians should particularly inquire about associated symptoms (*e.g.*, ocular or nasal symptoms, symptoms suggestive of systemic disease), history of sinus disease or surgery, facial trauma, history of obstruction of the nasolacrimal

drainage system, and quality of the tears (*e.g.*, tears tinted with blood could be a sign of neoplastic growth).³³ Patients with epiphora should have a full ocular examination and a fluorescein dye retention test with probing of the canaliculi and irrigation of the nasolacrimal system. Patients with concurrent nasal symptoms should undergo endoscopic examination of the nasal cavity to exclude an obstructive mass in the distal lacrimal system.^{33,34}

Identifying the specific or predominant cause of epiphora is essential for its proper management but can be challenging when epiphora is multifactorial, which is the case in $\sim 30\%$ of patients.³³ When epiphora is unilateral, excluding an obstructing neoplastic growth as a contributing factor of epiphora is critical.^{19,20} For example, in the present case, we described a patient who presented with bilateral lower lid ectropion and blepharitis, along with a small puncta on the side of the tumor, suggestive of punctal stenosis, all of which constitute potential independent risk factors for epiphora. Thorough history taking, which revealed epistaxis, and results of an examination, which demonstrated a total nasolacrimal duct obstruction, however, led to an endoscopic examination and then computed tomography, both of which showed the true cause of epiphora to be a neoplasm. By blocking the nasolacrimal duct, the neoplasm prevented appropriate tear drainage down the canaliculi and lacrimal sac, thereby resulting in epiphora.

EMC obstructing the nasolacrimal duct was ultimately found to be the predominant cause of our patient's epiphora. First described in 1972 by Donath *et al.*,³⁵ EMC is a rare low-grade adenocarcinoma that generally occurs in the salivary glands. Most cases of EMC originate from the parotid gland, with fewer cases occurring in the submandibular gland and the minor salivary glands.³⁶ Rarely, EMCs have also been described in the nasal cavity. A literature search of the previous cases of EMC of the nasal cavity indicated that EMC had thus far been described as presenting only with nasal symptoms (Table 1).^{24–31} The present case is unique in that the patient presented primarily with epiphora in addition to nasal symptoms.

EMCs tend to present as solitary tumors, and they can be easily diagnosed on biopsy by a combination of light microscopy and immunohistochemistry.^{23,36} A distinctive feature of EMC on light microscopy is a glandular structure characterized by a ductal lining, which consists of an inner layer of epithelial cells and an outer layer of clear myoepithelial cells.²³ Immunohistochemistry can then be used to definitely identify these two layers; for example, CAM 5.2 is a selective epithelial cell marker and α smooth muscle actin, p63, and calponin are selective myoepithelial markers.³⁶ Also, characteristic of EMCs is local invasion of tissues and a low-grade histology (*e.g.*, absence of nuclear

Table 1 Reported cases of epithelial myoepithelial carcinoma arising in the nasal cavity

Case Report	Site	Presentation
Harada <i>et al.</i> , ²⁴ 1996	Septum	"Nasal obstruction for several months and a bloody nasal drip for about 2 years"
Jin <i>et al.</i> , ²⁵ 1999	Septum	"A 2 months' history of progressive nasal obstruction"
Lee <i>et al.</i> , ²⁶ 2000	Inferior turbinate	"A 1-year history of nasal obstruction"
M'sakni <i>et al.</i> , ²⁷ 2007	Not specified	"Right recurrent epistaxis"
Yamanegi <i>et al.</i> , ²⁸ 2008	Inferior turbinate	"Recurrent epistaxis of the left nasal nostril of 3 months' duration"
Park <i>et al.</i> , ²⁹ 2011	Inferior turbinate	"Nasal obstruction with left recurrent epistaxis"
Ye and Guo, ³⁰ 2012	Not specified	Not specified
Chung <i>et al.</i> , ³¹ 2013	Not specified	"Bilateral nasal obstruction for several months"
Present case	Lateral nasal sidewall	"A 2-year history of epiphora in the left eye and a 1-year history of intermittent nasal crusting and bleeding"

pleomorphism, low mitotic rate),^{23,36} with rare exceptions in which EMCs can be highly aggressive and even lethal.^{29,36-39}

CONCLUSION

EMC is a rare salivary gland malignancy that can occur in the nasal cavity. It is a low-grade lesion but has metastatic potential, and early diagnosis and treatment are crucial for successful management. Unilateral epiphora with concurrent nasal symptoms should prompt a thorough evaluation of the nasal cavity to ensure that obstructive tumors are not present.

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