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Adenocarcinoma of the Ethmoid Sinus Presenting with Epiphora

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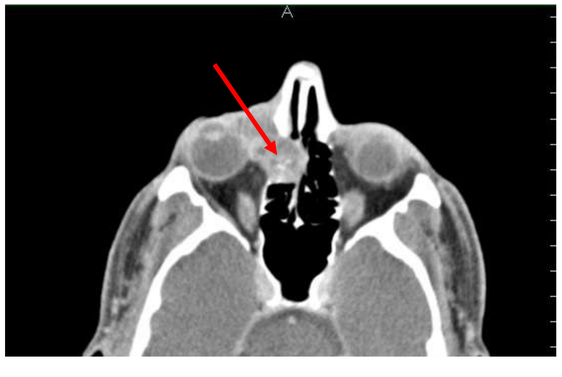
Introduction

- Neoplastic processes represent a rare but important etiology of acquired nasolacrimal duct obstruction. This includes primary tumors that arise within the lacrimal system as well as tumors in nearby tissues that secondarily compromise its drainage.
- Low-grade non-intestinal type adenocarcinomas (non-ITAC) are extremely rare tumors that arise from the surface epithelium of the sinonasal tract
- They frequently involve the ethmoid sinus, the nasal cavity, and the maxillary sinus
- These tumors present at a mean age range of 37-53 years with various non-specific symptoms including nasal obstruction and epistaxis
- The authors herein report the first case of ethmoidal low-grade non-ITAC presenting with symptoms of persistent unilateral epiphora. The clinicopathological findings and management of this patient are discussed.

Case Report

- A 63-year-old man first visited presented in May 2015 complaining of excessive tearing from the right eye that began four years earlier
- Examination revealed a non-tender, palpable mass in the right medial canthal (RMC) area that had not been noticed previously by the patient
- Probing and irrigation of the right lower lacrimal canaliculus revealed complete obstruction. Nasal endoscopy was performed and showed significant septal deviation to the right but no intranasal masses.
- Visual acuity, ocular motility, and the anterior segment were all within normal limits
- With a preliminary diagnosis of complete right sided nasolacrimal duct obstruction, endoscopic dacryocystorhinostomy (DCR) with placement of a nasolacrimal stent (Crawford tube) was recommended to the patient. He opted to wait until a later date to have the procedure done.

Imaging



- **Figure 1:** Axial computed tomography of the paranasal sinuses showing a large tumor mass in the ethmoid sinus invading into the right medial orbit

Case Report – Continued

- The patient returned in June 2019 complaining of progressive swelling and a hard mass in the RMC area associated with worsening epiphora and haemolacria
- Examination of the area revealed a small bump beneath the skin without overlying edema or erythema. Gentle palpation caused the mass to burst with a green mucopurulent discharge.
- CT scans of the head and orbits revealed a large growth in the ethmoid sinus invading into the right medial orbit and extending into the maxilla. [Fig. 1]
- Endoscopic endonasal biopsy of the mass was performed and a 1.4 x 1.2 x 0.2 cm section of the tumor mass was excised from the from the right ethmoid sinus
- Upon histopathological analysis, the tumor tissue demonstrated a cribriform pattern, moderately graded nuclei with occasional necrosis, and eosinophilic features [Fig. 2,3]
- Immunohistochemistry was performed, and the tumor cells stained positively for markers including androgen, GATA3, HER2/Neu. The tumor cells stained negative for CDX-2
- The tumor's location, morphology, and immunostaining patterns confirmed the diagnosis of low to intermediate grade non-intestinal type sinonasal adenocarcinoma of the ethmoid sinus.
- Several treatment options were discussed with the patient including surgical resection, chemotherapy, and radiation. Ultimately, the patient opted to undergo orbital exenteration and large sinus resection.

Histopathology

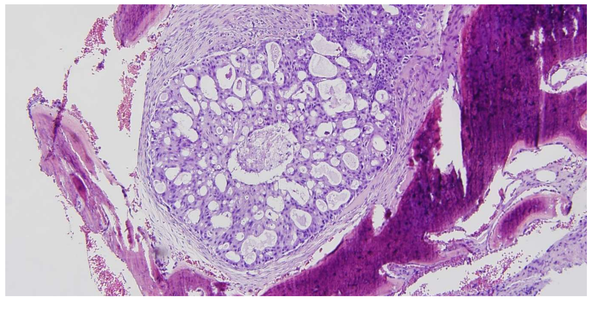
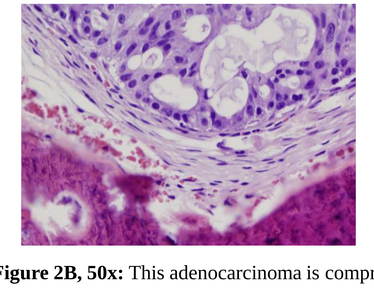


Figure 2A, 10x: Section from the mass of the right nasal passage and orbit showing adenocarcinoma with a cribriform pattern and surrounding bone (H&E 10x).



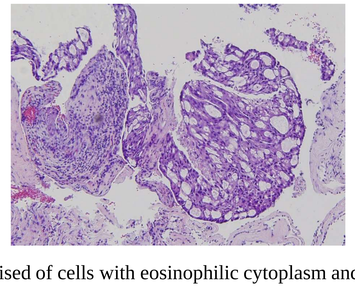


Figure 2B, 50x: This adenocarcinoma is comprised of cells with eosinophilic cytoplasm and moderate grade nuclear features (H&E 50x).
Figure 3, 10x: Section from the right middle meatus showing fragments of adenocarcinoma and background fibrous tissue and inflammation. (H&E 10x).

Laboratory Values and Printing Tips

- Cancers of the paranasal sinuses are extremely uncommon and account for just 1% of all human cancers (1)
- Synonyms for low-grade non-intestinal type adenocarcinoma in the literature include terminal tubulus adenocarcinoma,(2) sinonasal tubulopapillary low-grade adenocarcinoma,(3) and sinonasal seromucinous adenocarcinoma. (4)
- Microscopic analysis is key for differentiation of this tumor type from other similar sinonasal malignancies as well as for prognostic purposes
- Histologically, non-ITAC is recognized by its trabecular, cribaform, or papillary growth pattern and the single layer of uniform columnar or cuboidal cells that line its glands. (5) Pleomorphic nuclei and mitotic figures are rarely seen in this subtype.
- Immunohistochemically, non-ITAC is characterized by positive CK7 staining and/or negative CDX-2 and CK-20 staining. (6)
- Differential diagnoses to consider include intestinal type adenocarcinoma (ITAC), acinic cell carcinoma, and oncocytic Schneiderian papilloma.
- ITAC is an aggressive malignancy differentiated from non-ITAC by its resemblance to the mucosa found in the small and large intestine, its high-grade histologic appearance, and cellular expression of CDX-2.
- Distinguishing between the two tumors types is of crucial clinical importance given that low-grade non-ITAC has not, to date, displayed metastatic behavior while several instances of low-grade ITAC metastases have been reported (7)
- Rarely, non-intestinal type adenocarcinoma may be confused with oncocytic Schneiderian papillomas. However, the epithelium of oncocytic Schneiderian papillomas are multilayered and lack glandular lumina. (8)
- The presenting clinical symptoms of non-ITAC vary, however the most common reported symptoms are nasal obstruction and epistaxis (9)
- In our case, the patient presented with persistent unilateral epiphora which has not, to date, been reported in association with this tumor subtype.
- The mainstay in treatment of low-grade non-ITAC includes surgery followed by radiation therapy.
- Overall, the prognosis of patients with non-ITAC is good and rarely does death occur due to the disease.

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