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Case Report

Primary Orbital Mucinous Carcinoma in a Human Immunodeficiency Virus (HIV) Patient: A Case Report

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Abstract

Mucinous carcinoma is a rare malignancy marked by abundant mucin production, typically found in organs like the breast, colon, and ovary, with orbital involvement being exceptionally rare. This case report presents a 55-year-old male with a history of well-controlled Human Immunodeficiency Virus (HIV) infection, oral squamous cell carcinoma, and recurrent papillomas of the conjunctiva and oropharynx, who developed a firm, enlarging mass in the left superomedial anterior orbit. Histopathology after surgical resection confirmed mucinous carcinoma, and full-body imaging ruled out metastasis, but demonstrated an intensely hypermetabolic left lacrimal sac fossa/upper nasolacrimal duct on Positron Emission Tomography (PET) imaging. Although a maxillectomy and post-excision radiation was advised, the patient declined further intervention. This report highlights the importance of ongoing surveillance in immunocompromised patients, as even those adherent with treatment may develop rare and aggressive malignancies. While Human Papillomavirus (HPV) has known oncogenic potential, its role in orbital mucinous carcinoma remains unclear, warranting further investigation.

Keywords: Orbital Mucinous Carcinoma; Human Immunodeficiency Virus

Introduction

Mucinous carcinoma is a rare malignant epithelial tumor characterized by the production of abundant extracellular mucin. While it is most frequently observed in the breast, colon, and ovary, cutaneous variants may also arise in the head and neck region, particularly in the periocular area. Primary mucinous carcinoma of the orbit is exceedingly rare, with only one true case documented in the literature to date [1]. These types of tumors are typically indolent but are known for their potential for local recurrence and distant metastasis [1]. The rarity of this tumor in the orbit poses challenges in both diagnosis and management, especially in patients with complex medical histories. Immunocompromised individuals, such as those living with Human Immunodeficiency Virus (HIV), are at increased risk for both typical and atypical malignancies [2,3]. Additionally, the oncogenic properties of Human Papillomavirus (HPV) have been implicated in various epithelial cancers, including those arising in the conjunctiva and oropharynx [4]. To date, there is no known association between HPV-related papillomatous disease and mucinous carcinoma. In this report, we present a unique case of primary orbital mucinous carcinoma in an HIV-positive patient with a history of recurrent intranasal papillomatous lesions, highlighting the diagnostic and clinical challenges posed by this rare presentation and underscoring the importance of surveillance in high-risk populations. The collection and evaluation of protected patient health information was Health Insurance Portability and Accountability Act compliant during the preparation of this manuscript. This report adheres to the ethical principles outlined in the Declaration of Helsinki.

Case Presentation

A 55-year-old male with known HIV infection (CD4 count 501, undetectable HIV RNA) and a remote history of left conjunctival papilloma was found to have multiple papillomatous lesions in the oropharynx, nasal passageways, and left nasolacrimal duct. Biopsies confirmed HPV-positive invasive squamous cell carcinoma of the left larynx, and he subsequently underwent chemotherapy and external beam radiation. Several months later, he developed left dacryocystitis and underwent an external Dacryocystorhinostomy (DCR). Intraoperatively, a papillomatous lesion was again identified in the lacrimal sac, and biopsy of the lesion and of additional nasal papillomas were negative for malignancy. Seven months later, he developed a firm, non-tender palpable mass in the left medial upper eyelid, with imaging showing a lesion in the superomedial anterior orbit (Fig. 1). Visual acuity, intraocular pressure, extraocular motility, and pupillary responses were within normal limits. Histopathologic examination of the specimen demonstrated islands of basoloid epithelial islands within abundant pools of mucin material. The tumor cells demonstrated positive reactivity with Cytokeratin (CK)-5/6 and CK-7 and negative reactivity with CK-20 and CDX2. The overall features were diagnostic of mucinous carcinoma of the orbit. A comprehensive metastatic work-up, including whole-body Positron Emission Tomography (PET) imaging, revealed an intensely hypermetabolic left lacrimal sac fossa/upper nasolacrimal duct with no evidence of metastatic disease or other primary site. Despite tumor board recommendations for maxillectomy followed by adjuvant radiation, the patient declined further surgical intervention and remains under close clinical surveillance (Fig. 2,3).

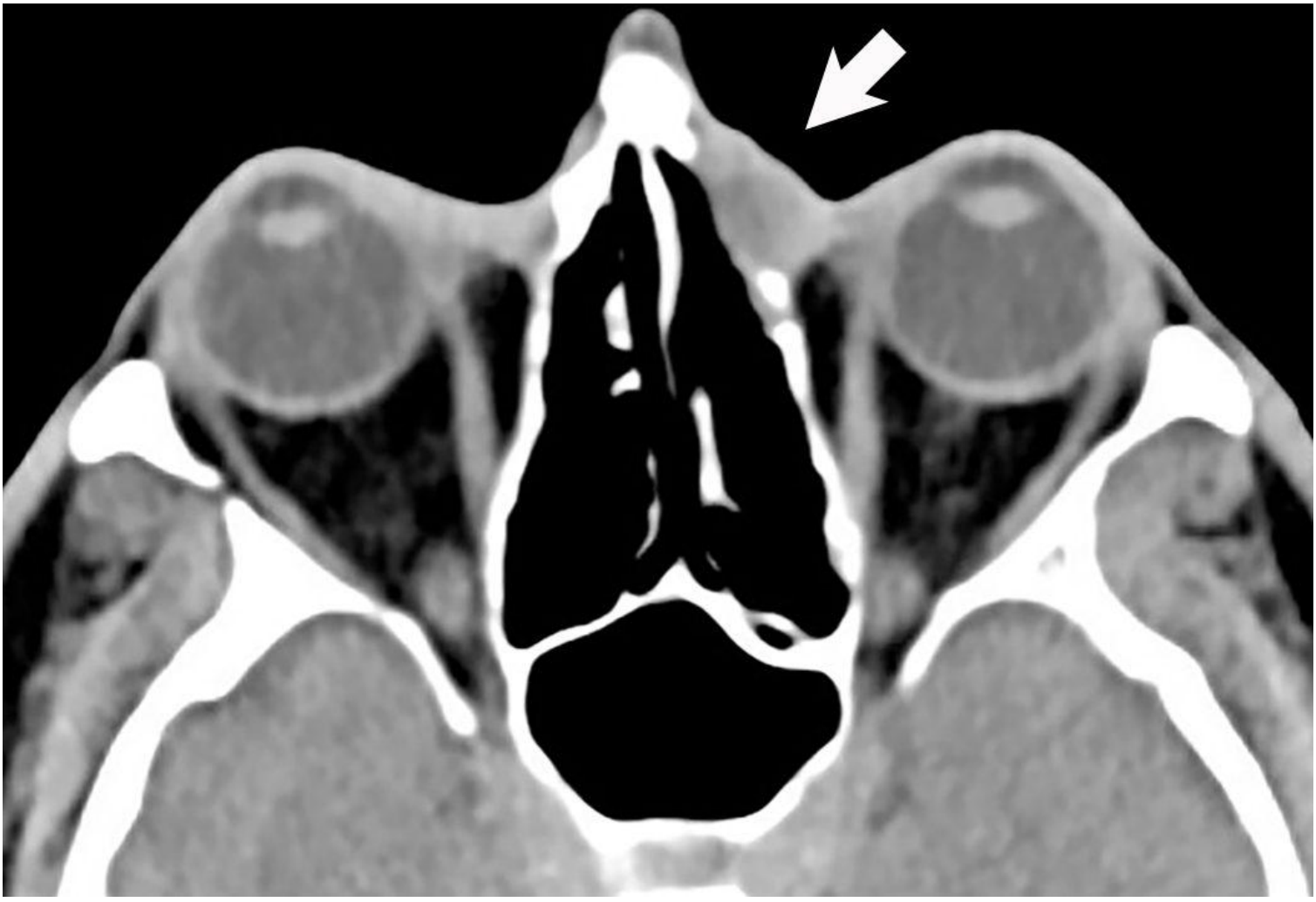

Figure 1: Computed Tomography (CT) scan with axial view of the orbits demonstrating a well-defined, soft tissue mass along the left medial upper eyelid with extension into the anterior orbit (arrow).

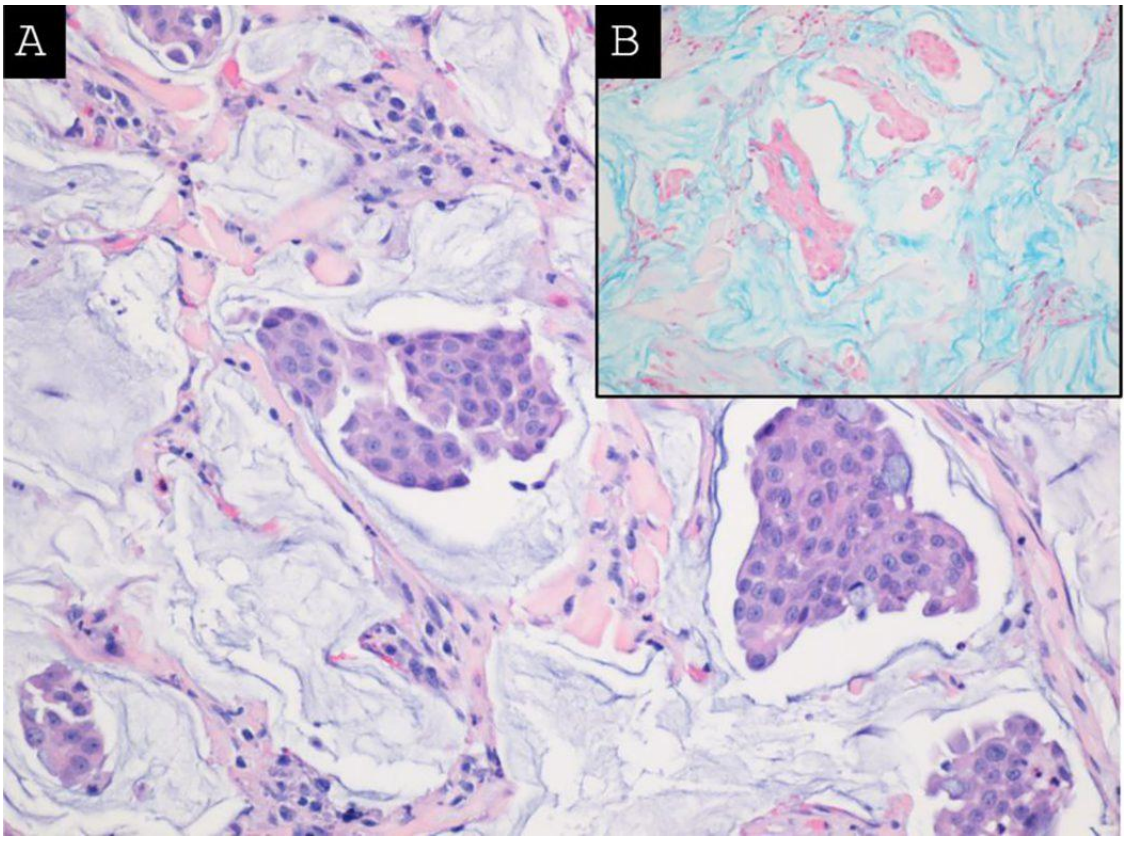

Figure 2: A. Histologic appearance of orbital tumor showing basoloid epithelial islands within pools of mucin material (hematoxylin and eosin stain, original magnification x 200). B. Alcian blue stain pH 2.5 demonstrating abundant mucin (original magnification x 200).

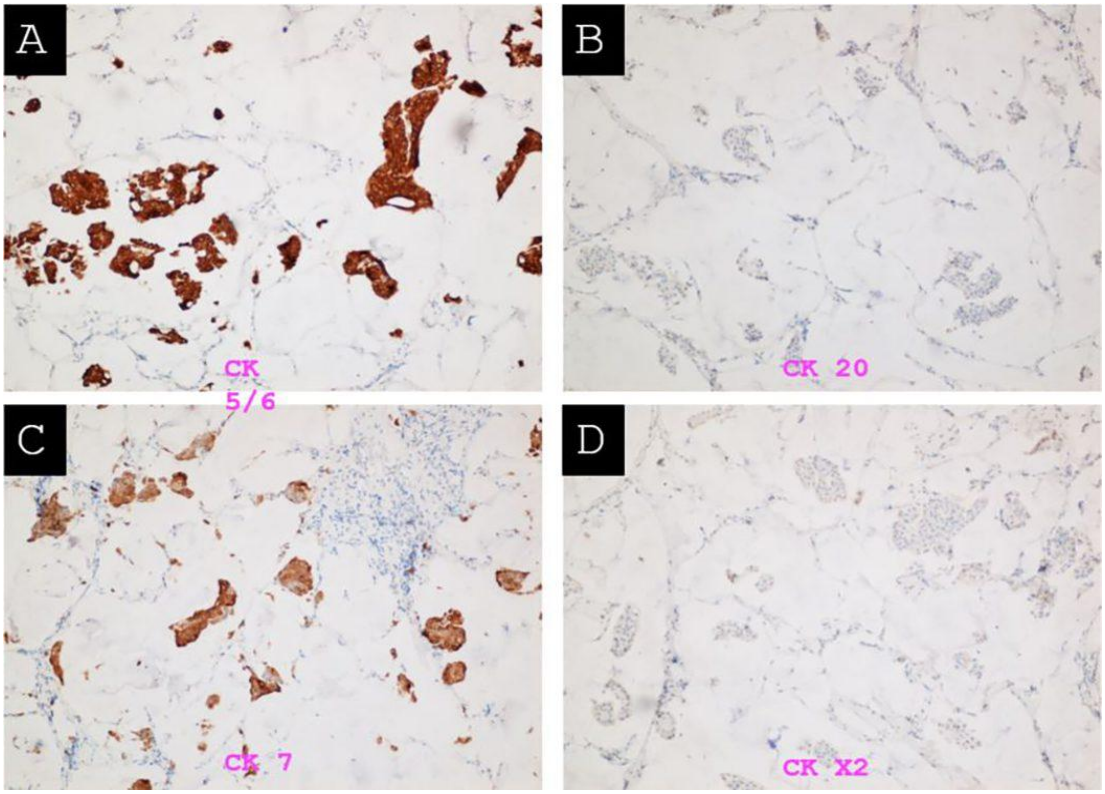

Figure 3: Immunohistochemical studies (A) cytokeratin (CK)-5/6 positive (B) CK-20 negative (C) CK-7 positive (D) CDX2 negative (original magnification x100).

Discussion

Primary mucinous carcinoma of the orbit is an exceptionally rare malignancy, with only one case reported in the literature [1]. These tumors are histologically characterized by clusters of epithelial cells floating in pools of extracellular mucin and can be difficult to distinguish from metastatic mucinous carcinomas originating from distant sites [1,5]. Thorough metastatic workup is critical to confirm a primary orbital origin. While there is a lack of literature regarding orbital mucinous carcinoma, previous reports of eyelid mucinous carcinoma, have emphasized the diagnostic importance of ruling out systemic disease [6]. In our patient, a thorough evaluation revealed no evidence of other primary sites or metastasis, supporting the diagnosis as a primary orbital mucinous carcinoma. While there was a prior history of external DCR, and it is possible that this lesion arose from the lacrimal sac mucosa or adjacent eyelid, the lack of any cutaneous or intranasal involvement supports a primary orbital origin of the pathology. Immunocompromised patients, particularly those with HIV, are at increased risk for both infectious and neoplastic diseases, including rare malignancies [2-5]. Although this patient had well-controlled HIV with adherence to antiretroviral therapy, his history of HPV-associated laryngeal squamous cell carcinoma and recurrent conjunctival and oropharyngeal papillomas adds an additional layer of oncologic complexity. While HPV has a well-documented role in epithelial carcinogenesis, especially in the conjunctiva and upper aerodigestive tract, no link between HPV and mucinous carcinoma of the orbit has been established [6,7]. This case raises the possibility of a shared oncogenic environment in immunocompromised patients with chronic HPV-related lesions.

Management of primary mucinous carcinoma in the periocular and orbital regions is not standardized due to its rarity. In our case, tumor board recommended maxillectomy and adjuvant radiation. However, the patient declined further surgical intervention, emphasizing the importance of shared decision-making and the need for vigilant follow-up in cases where definitive therapy is deferred.

Conclusion

This case also underscores the necessity of maintaining a high index of suspicion for rare malignancies in immunocompromised patients. While mucinous carcinoma is rare in the orbit, this report highlights the need for further studies to determine any potential etiologic role of HPV or chronic mucosal papillomatosis in its pathogenesis. Close, long-term surveillance is essential in managing patients at high risk for atypical malignancies.

Conflict of Interest

The authors declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

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Data Availability Statement

The data supporting the findings of this study are available from the corresponding author upon reasonable request.

Ethical Statement

The project did not meet the definition of human subject research under the preview of the IRB according to federal regulations, and therefore was exempt.

Informed Consent Statement

Informed consent was obtained from all participants included in the study.

Authors' Contributions

All authors contributed equally to this paper.

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