Mucoepidermoid carcinoma ex-inverted papilloma

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Abstract

Mucoepidermoid carcinoma (MEC) typically arises from major salivary glands, minor salivary glands of the oral cavity and mucous glands of the aerodigestive tract. Cases of MEC arising from benign sinonasal or lacrimal papillomas are extremely rare. Herein we describe a MEC representing malignant transformation of an inverted Schneiderian papilloma. The patient presented with nasal bridge pain radiating to the premaxillary regions. Imaging showed a tumour involving the left nasal cavity, left ethmoid and maxillary sinuses and extending into the left orbit and lacrimal sac. Initial biopsy and debulking specimens showed only inverted papilloma. The re-excision specimen demonstrated a tumour with features of both inverted papilloma and MEC that superficially invaded bone. The tumour was negative for MECT1-MAML2 translocation. Linear array analysis of tumour tissue demonstrated the presence of human papillomavirus-11 (HPV-11).

Keywords inverted Schneiderian papilloma; lacrimal system; mucoepidermoid carcinoma; sinonasal tract

Introduction

Schneiderian mucosa lines most of the paranasal sinuses and the nasal cavity with the exception of the nasal vestibule and the roof of the nose, which are lined by stratified squamous epithelium and olfactory mucosa respectively. Three benign neoplastic papillomatous neoplasms arise from the Schneiderian membrane: exophytic (also referred to as fungiform or everted) papilloma, inverted (endophytic) papilloma, and oncocytic (cylindrical cell) papilloma.[1] Transformation to squamous cell carcinomas has been reported to occur in up to 2% of sinonasal papillomas,[2,3] with the vast majority of such cases occurring in inverted papillomas. Transformation to other types of carcinomas is much less frequent with reported cases including sinonasal type undifferentiated carcinoma, mucoepidermoid carcinoma, adenocarcinoma NOS, and small cell neuroendocrine carcinoma.[1,2,3]

Case report

The patient is a 43-year old woman who presented to the ocular clinic with a one-year history of pain over the bridge of the nose which radiated bilaterally to the premaxillary region. She denied any ocular symptoms or epistaxis, although she reported coughing up nasal secretions mixed with blood on several occasions. Her past medical history included hepatitis C, intravenous drug use, back trauma and ovarian cystectomy. MR imaging at presentation showed a large nasal cavity mass extending into the anterior left ethmoid air cells, the left nasolacrimal canal and lacrimal sac, and through the left lacrimal bone into the orbit. The signal characteristics were suggestive of a cellular tumour such as inverted papilloma or carcinoma. Transnasal debulking of the nasal portion of the lesion was performed and the surgical specimen showed inverted Schneiderian papilloma without malignant transformation.

The patient's symptoms persisted after her initial surgery and a follow-up CT scan showed residual tumour in the left ethmoid and the maxillary sinuses, the left lacrimal sac as well as the left medial orbit (Figure 1). The orbital portion of the tumour was resected at a different institution through a left posterior orbitotomy and medial canthopexy. The pathology specimen from that resection showed papillomatous light pink tumour fragments of inverted papilloma with numerous mucous cells. Re-excision of the sinonasal mass was performed via external-approach rhinotomy, left medial maxillectomy, left sphenoidectomy and ethmoidectomy. The specimen resected during this last surgical resection was reported as "low-grade mucoepidermoid carcinoma ex-inverted papilloma". The patient underwent adjuvant radiotherapy and is well with no tumour recurrence 9 months after her last surgery.

Gross examination showed papillary pink, tan, and white tissue fragments admixed with membranous mucosal and bone fragments. Microscopic examination showed the lesion consisting predominantly of a thickened, multilayered non-keratinizing squamous epithelium replacing normal respiratory mucosa, and seromucous glands and ducts. The neoplastic cells showed an inverted or endophytic growth pattern extending into the underlying stroma consistent with inverted Schneiderian papilloma (Figure 2a); however, admixed with the squamous epithelium were a large number of mucous and goblet cells (Figure 2b), more than would be expected in an inverted papilloma. Additionally, multiple foci of cytologic atypia, nuclear pleomorphism (Figure 2c) and invasion of the bone were noted (Figure 2d). These microscopic findings, taken together with the aggressive nature of the tumour and its high rate of growth, caused the

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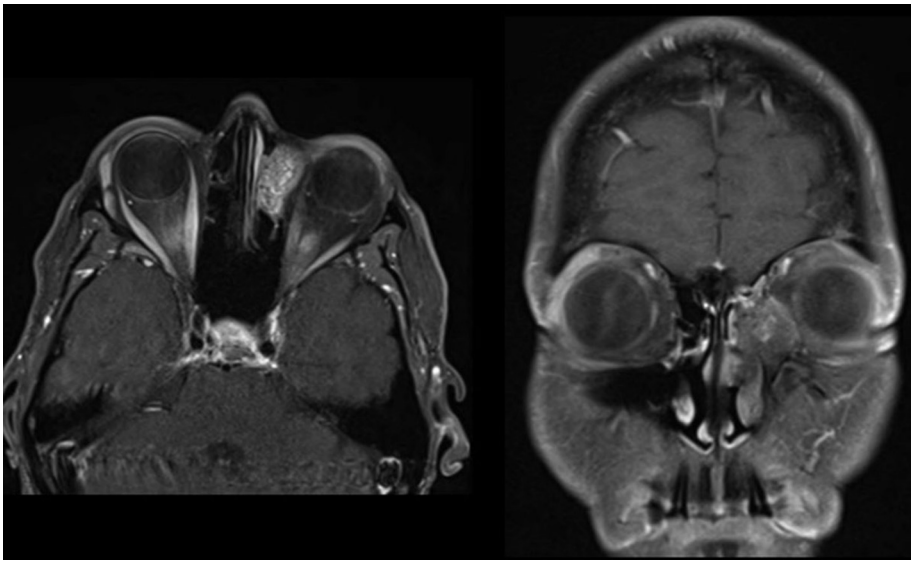

Figure 1 Axial and coronal T1 weighted post gadolinium enhanced images demonstrates mild enhancement of the left anterior ethmoid region mass that has extended into and is enlarging the nasolacrimal sac fossa.

classification of this tumour as a low grade mucoepidermoid carcinoma arising from an inverted Schneiderian papilloma (i.e. a low grade mucoepidermoid carcinoma ex-inverted Schneiderian papilloma).

Immunohistochemistry for p16 showed strong but heterogeneous staining in about 30% of the tumour cells (Figure 3a). Ki-67 labeling index was approximately 20% (Figure 3b). HPV molecular testing by Linear Array demonstrated the presence of

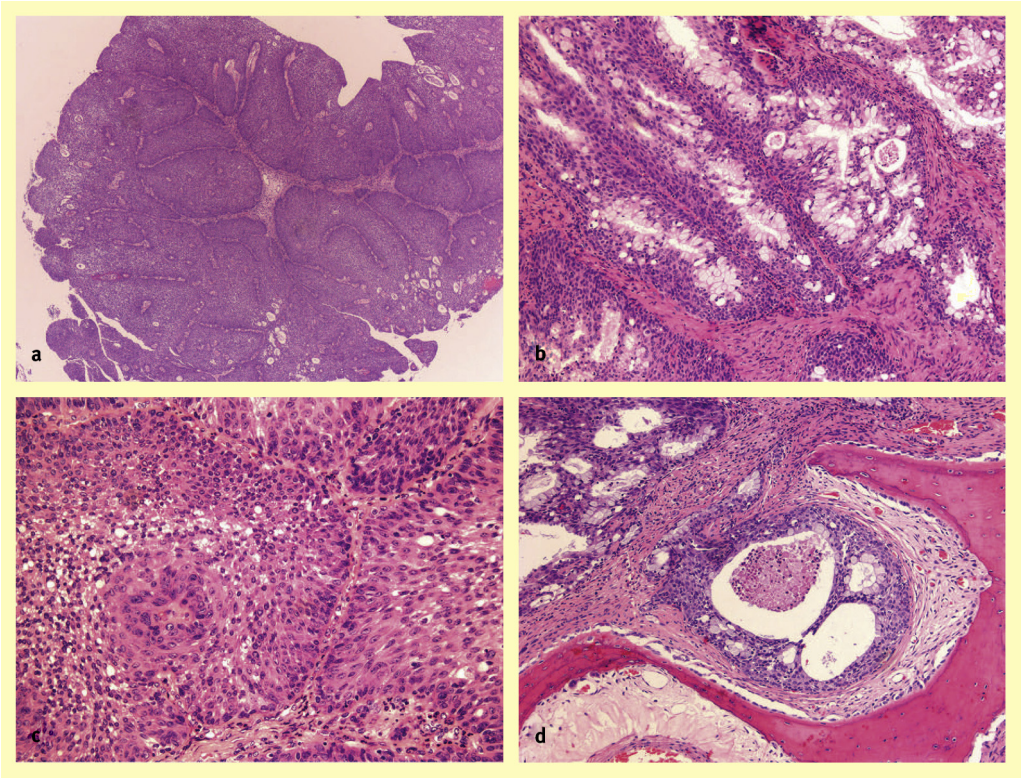

Figure 2 Papillomatous area composed of non-keratinizing squamous epithelium with an endophytic grow pattern (**a**). Invasive carcinoma composed of atypical non-keratinizing squamous cells and numerous mucous goblet cells (**b**). Atypical squamous cells showing hyperchromatic and pleomorphic nuclei (**c**). Focus of mucoepidermoid carcinoma close to trabeculae of ethmoid bone (**d**).