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Case Discussion

# Endoscopic trans-pterygoid resection of a low-grade cribriform cystadenocarcinoma of the infratemporal fossa

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**Abstract** This article presents a case of low-grade cribriform cystadenocarcinomas (LGCCC), a rare salivary gland tumor manifesting in the infratemporal fossa (ITF). The lesion in this case is unique in its location, histopathology, and management in that the tumor resection was performed using an exclusively endoscopic, endonasal approach. This case highlights the expanding application of endoscopic skull base techniques to address an indolent, slow-growing malignancy of the ITF.


## Introduction

Low-grade cribriform cystadenocarcinoma (LGCCC) is a recently described rare salivary gland neoplasm that largely arises in the parotid gland and exhibits an indolent course. In 1996, Delgado et al[1] originally defined this tumor as a low-grade salivary duct carcinoma (LGSDC). In the third edition of the World Health Organization (WHO) Classification of Head and Neck Tumors, it was initially categorized as a variant of cystadenocarcinoma. However, the most

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recent edition of the WHO Classification has reclassified this entity as a low-grade intraductal carcinoma.[2] As a rule, LGCCC is a parotid tumor but rare cases have been reported to arise from the submandibular gland, accessory parotid gland, infra-parotid lymph nodes, and palate.[3–6] In this report, we detail the only case in the English literature of primary LGCCC arising from tissue within the infratemporal fossa (ITF) and its surgical management using an exclusively endoscopic, endonasal approach.

## Case description

A 46-year-old woman presented with an incidental finding of an ITF mass on magnetic resonance imaging (MRI) that was performed in preparation for spinal surgery. She complained of mild right-sided facial pain and headache, but was otherwise asymptomatic. Physical examination and sinonasal endoscopy were unremarkable. An MRI of the head and neck revealed a well-circumscribed, lobulated mass of the pre-styloid parapharyngeal space, measuring 2.5 cm $\times$ 2 cm (Fig. 1). Computed tomography angiography (CTA) demonstrated an enhancing mass that displaced the right internal carotid artery posteriorly without invasion, and branches of the right external carotid artery coursing along its lateral and postero-lateral margins. A benign tumor or low-grade malignancy of the parapharyngeal space was suspected. Surgery was offered, including endoscopic medial maxillectomy and trans-pterygoid approach under stereotactic, computer-assisted navigation.

A nasoseptal flap was elevated at the outset of the case in preparation of reconstruction of the ITF defect. A posterior septectomy was performed, providing binarial access. A modified Denker's approach, transecting the nasolacrimal duct, and medial maxillectomy were then performed. A trans-pterygoid approach to the infratemporal fossa was then performed using a 2-surgeon, 3-handed technique. The tumor was visualized and found to be well-encapsulated. The lesion was then dissected circumferentially from its soft tissue attachments and removed en bloc. Because of the large size of the defect and its proximity to the internal carotid artery, an abdominal fat graft was harvested and used in conjunction with the nasoseptal flap to reconstruct the ITF defect.

The histopathologic and immunohistochemistry studies identified the tumor as a low-grade cribriform cystadenocarcinoma, likely arising from the deep lobe of the parotid gland. The lesion was resected in its entirety, with clear tumor margins. At 8 months postoperatively, follow-up imaging, including MRI and positron emission tomography/computed tomography (PET/CT), showed no sign of recurrent disease or distant metastases.

## Discussion

LGCCC is a rare neoplasm that has recently been reclassified by the WHO as a low-grade intraductal carcinoma.[2] Originally, the malignancy was considered to be a low-grade variant of salivary ductal carcinoma. However, both its clinical behavior and histopathologic features have shown that the tumor differs from other salivary duct carcinomas in its pattern of growth and the absence of pathologic characteristics such as nuclear atypia, invasiveness into surrounding tissue, or metastasis to regional lymph nodes. Since its initial description in 1996, 47 cases have been described in the literature, including the present case. Although the number of cases with long-term follow up is low, none have shown evidence of recurrence, metastasis, or mortality from the tumor.[7–9] The majority of tumors has arisen within the parotid gland, but has also infrequently involved the submandibular gland, palate and minor salivary glands. To the best of our knowledge, there have been no reported cases of LGCCC (or LGSDC) arising in the infratemporal fossa.

There have been only been a handful of reports describing the morphological findings of LGCCC. The findings included: (1) proliferation of ductal epithelial cells with tight junctions; (2) mild nuclear atypia and minimal size variation; and (3) cytoplasmic vacuoles and squamoid or metaplastic changes in tumor cells.[7,9] The histology in this case revealed neoplastic epithelial cells with moderate atypia and nuclei that lacked any significant atypia or size variation. The tumor cells formed glands that varied significantly in size. In the larger glands, tumor cells demonstrated a cribriform growth pattern and signs of mucin production (Fig. 2). Moreover, there was no evidence of tumor invasion into the surrounding structures, tumor

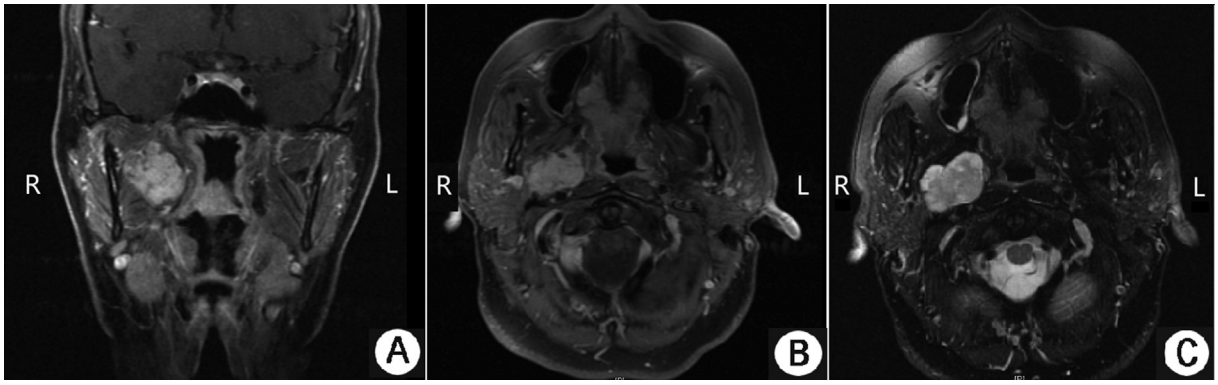


**Figure 1** A: Coronal T1-weighted MRI sequence demonstrating a lobulated, well circumscribed mass in the right infratemporal fossa. B: Axial T1 MRI demonstrating a T1-weighted hypoerintense lesion of the infratemporal fossa. C: Axial T2 MRI showing this lesion with T2-weighted hyperintensity.

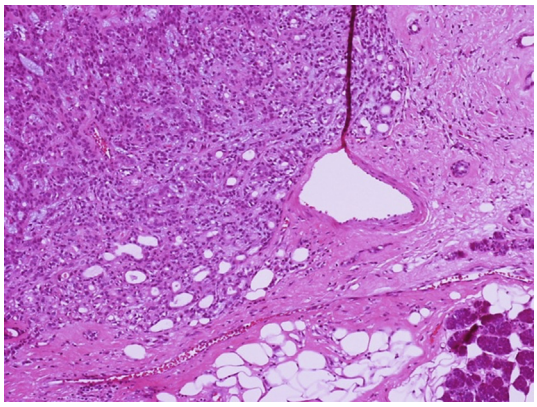

**Figure 2** Neoplastic epithelial cells with moderate atypia with nuclei of uniform size and shape. Larger cystic areas show tumor cells in a typical cribriform growth pattern.

necrosis, perineural extension, or vascular invasion, similar to other published reports of LGCCC. Immunohistochemistry revealed strong, diffuse, S100 positivity and the presence of myoepithelial markers (e.g., calponin) surrounding cystic spaces — consistent with previously reported cases of this tumor type.

Due to the favorable outcome of the diagnosis of LGCCC, a fine-needle aspiration (FNA) of the tumor is usually desirable for initial histopathologic examination.[10] That was not possible in this particular case due to the location of the tumor within the ITF. Thus, the decision was made to proceed with an operative excisional biopsy for frozen histopathologic analysis and complete surgical removal of the tumor.

With respect to treatment, this case highlights the feasibility of complete surgical resection with an endoscopic, endonasal approach to the ITF. This approach provided a shorter recovery time, improved cosmesis, and reduced morbidity in comparison to more traditional subtemporal, transtemporal and anterior transfacial approaches. We consider this a reasonable application of an endoscopic skull base technique to address an indolent, slow-growing malignancy of the ITF.

## Declaration of Competing Interest

Kibwei A. McKinney is a part of the Speakers Bureau for Intersect ENT.

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