Intracranial Salivary Gland Choristoma within Optic Nerve Dural Sheath: Case Report and Review of the Literature

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Key words
- Choristoma
- Ear, middle
- Optic nerve
- Salivary glands
- Sella turcica

Abbreviations and Acronyms
MRI: Magnetic resonance imaging

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OBJECTIVE: **Salivary gland choristomas or their neoplastic derivatives may appear throughout the intracranial space, most frequently in the middle ear or sella. Here, we present the case of a salivary gland choristoma embedded within the optic nerve dural sheath and review the literature of intracranial salivary gland masses.**

CASE PRESENTATION: **A 28-year-old female patient presented with headache and visual complaints. Magnetic resonance imaging revealed a prechiasmatic suprasellar cystic lesion. Operatively, the mass appeared as a cyst filled with mucinous fluid associated with abnormal tissue embedded within the optic nerve.**

RESULTS: **We deflated and biopsied the cyst, which revealed normal-appearing salivary tissue. The patient remains asymptomatic without radiographic evidence of cyst recurrence 2 years postoperatively.**

CONCLUSION: **Intracranial salivary tissue has been previously described but never before in the suprasellar space. Although rare, knowledge of their natural history and pathologic features may inform surgical management if they are encountered in the operating room.**

INTRODUCTION

Salivary gland tissue is typically confined to the major (sublingual, submandibular, and parotid) and minor salivary glands. Although islands of salivary tissue have been reported in sites as distant as the prostate and perianal region, choristomas typically are found within the head and neck region (5). Intracranial cases have been previously reported; however, here we discuss a unique presentation of patient with a suprasellar salivary gland choristoma.

CASE PRESENTATION

The patient, a 28-year-old woman, presented with a 1-month history of intermittent headaches and progressively blurred vision in the left eye that had precipitously decreased in the week before her evaluation. She also complained of 2 weeks of insatiable appetite with a 15-pound weight gain during the same period, as well as frequent urination. Her primary care physician referred her for a computed tomography scan, and she was instructed to present to the emergency department after the community radiologist appreciated a sellar-region mass. Except for slight weakness in her left extremities, her only physical examination abnormality was blurred vision in the left eye with decreased temporal field perception. A neuro-ophthalmologic examination confirmed decreased acuity; however, visual field testing was not completed because perimetry testing made the patient intolerably nauseated. Magnetic resonance imaging (MRI), shown in Figure 1, demonstrated a nonenhancing suprasellar cyst that appeared hypointense on T1- and hyperintense on T2-weighted scans.

The patient was brought to the operating room during the same admission for a right subfrontal craniotomy. Once encountered, the cyst was fenestrated and slightly mucinous fluid was expelled with gentle pressure on the cyst wall. After the cyst was deflated, a small piece of yellow-tinged tissue heavily invested in the medial edge of the left optic nerve sheath was appreciated. The nerve itself appeared slightly wider than the contralateral nerve. A portion of this tissue was dissected and found to be diagnostic by frozen section. Further dissection was attempted but quickly abandoned out of concern for iatrogenic injury to the optic nerve. The remainder of the case was uneventful.

Microscopic examination (Figures 2 and 3) revealed branching tubules and small glands or acini lined by attenuated to columnar epithelium. Cells showed round regular nuclei and clear vacuolated cytoplasm. Lumens contained pale blue mucinous material. Secretory granules were not present. There were no respiratory or ciliated epithelium and no goblet cells. Atypia and mitotic activity were absent. The findings were benign and closely resembled mucous-type salivary tissue, similar to sublingual gland and accessory salivary glands, such as those found along the respiratory tract.

Postoperatively, the patient developed a number of sensorimotor complaints that, after thorough work-up, were thought to be the result of conversion disorder. She also endorsed nonspecific visual complaints, yet findings of a neuro-ophthalmologic

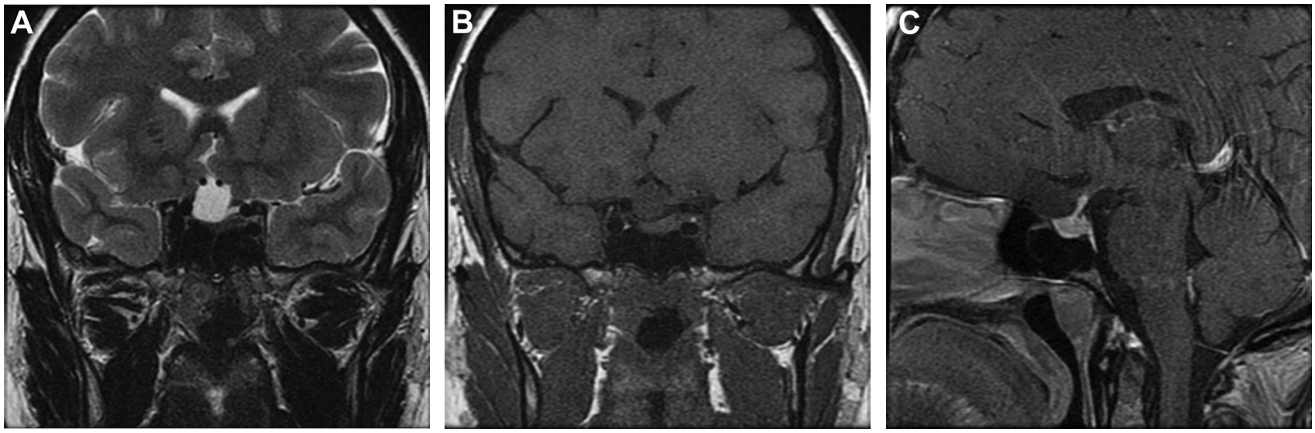

Figure 1. Coronal T1 (**A**) and T2 (**B**) magnetic resonance imaging demonstrating the homogeneous appearance of cyst beneath paired anterior cerebral arteries. A contrast-enhanced mid-sagittal T1 (**C**) shows the cyst in relation to sella and pituitary gland.

examination were normal. At her most recent visit almost 2 years after surgery, many of her nonorganic symptoms had improved. Her postoperative MRI findings have been stable and without suggestion of cyst recurrence (Figure 4).

DISCUSSION

Frequent locations of ectopic salivary gland tissue include the neck, gingiva, and middle ear (5). Middle-ear salivary gland choristomas are thought to be the result of developmental errors within the second, or less frequently first, branchial arch. Consequentially, they are commonly associated with a number of developmental abnormalities characteristic of an as-yet-unnamed syndrome (1). Salivary choristomas of the middle ear most frequently present with early-onset unilateral hearing loss. Because they tend not to grow and hearing loss generally remains stable, complete resection of these tumors is not recommended (1). Interestingly, a number of these choristomas have been noted to be intimately associated with the facial nerve (13), which may help explain the 12% incidence of iatrogenic facial nerve injury during surgical treatment (1).

A less frequent, although well-described, location of heterotopic salivary gland tissue is within the sella. These lesions may appear as either choristomas (2, 10, 11, 17) or neoplasm of salivary gland origin (7, 8, 15, 18). Both entities can be explained embryologically. During fetal development, the Rathke pouch—which is destined to become the anterior pituitary—initially forms as a depression in the mucosal roof of the mouth that grows in a cephalad direction to meet the down-growing projection of the diencephalon, which is destined to become the posterior pituitary. Either through direct transfer of intact seromucous glands from the oral cavity or induction of pituitary epithelium by salivary gland mesenchyme (8), microscopic salivary gland rests frequently can be demonstrated within normal pituitary glands (16). Perhaps because of the presence of oncocytes (16), these rests are at risk for malignant transformation in the same way as the major or minor salivary glands (8).

Intrasellar salivary choristomas may present with hormonal imbalances, whereas those with suprasellar extension may cause headache or visual changes from chiasmatic compression. Sellar neoplasms of salivary gland origin may present with the same, but a number have also caused extraocular muscle palsy, presumably from cavernous sinus invasion. Choristomas have generally been resected in gross-total fashion via a transsphenoidal route with postoperative symptomatic resolution. The neoplastic lesions have been approached from both transsphenoidal and transcranial routes for both gross total and subtotal resections. Long-term follow-up in these reports is lacking, but of the eight reported patients in this group, two died in the days after surgery: one from severe hypotension (7) and the other from rupture of a previously known thoracic aortic aneurysm (18). Four received postoperative radiation (8, 15), although in one case (15) it was administered after a biopsy was incorrectly assessed to be craniopharyngioma. In this case, the tumor recurred 14 years later and was resected.

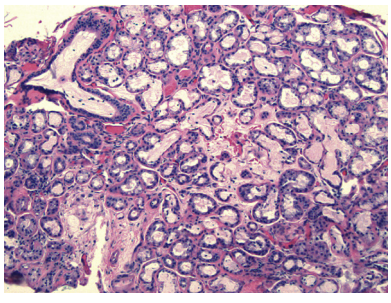

Figure 2. Hematoxylin and eosin stain at 10× objective. Branching tubules and acini lined with non-ciliated cuboidal epithelium.

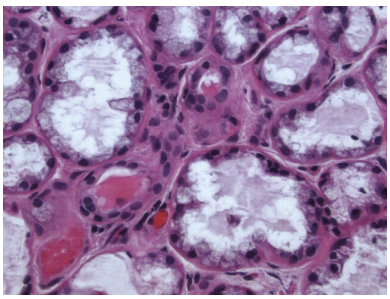

Figure 3. Hematoxylin and eosin stain at 40× objective. Note uniform nuclei arranged in an orderly fashion with mucinous material within the lumina.