Nasal Septal Adenoid Cystic Carcinoma: Case Report and Review of the Literature

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INTRODUCTION

ABSTRACT

Educational Objectives: At the conclusion of this presentation, the participants should be able to discuss the presentation and diagnostic work up for adenoid cystic carcinoma; and 2) be able to discuss the unique and distinguishing features of adenoid cystic carcinoma.

Study Design: Case report and review of the primary literature.

Methods: We present a case of a 70-year-old female with complaints of blurred vision for several months. Patient denied any history of epistaxis or nasal obstruction. An MRI was performed at an outside hospital showing a 1.5 cm lesion on the middle superior septum. Nasal endoscopy revealed a spherical enlargement of the middle superior septum primarily on the right. A biopsy was performed and ACC was diagnosed. A PET CT obtained was pertinent for a localized increased FDG uptake (SUV max 3.7) in the anterior nasal septum measuring 1.8cm. Considering these findings the patient underwent a right lateral rhinotomy and subtotal septectomy with the recommendation to receive postoperative radiation therapy.

Results: The surgical specimen was consistent with adenoid cystic carcinoma without bony or cartilaginous invasion and revealed clear margins. Review of the literature revealed that this entity is exceedingly rare, with only 3 described cases reported in the English literature, and the only one that presented with visual disturbances.

Conclusions: Adenoid cystic carcinoma of the septum is a rare entity, as discovered in our literature search. Early diagnosis is key for curative treatment so it should be considered in the differential diagnosis of any nasal septal mass. Treatment options should include wide local excision via an open or endoscopic approach and postoperative radiation treatment. Due to the high incidence of local recurrence and distant spread these patients require long-term follow up.

CASE REPORT

We present a case of a 70-year-old female with complaints of blurred vision for several months. An MRI of the orbits was performed at an outside hospital showing an incidental 1.5 cm soft tissue lesion on the middle superior septum. (see figure 1 for CT scan) Nasal endoscopy revealed a spherical enlargement of the middle superior septum primarily on the right. A biopsy was performed and ACC was diagnosed. A PET CT obtained was pertinent for a localized increased FDG uptake (SUV max 3.7) in the anterior nasal septum measuring 1.8 cm. There was no evidence of FDG avid locoregional or distant metastasis.

The patient underwent a right lateral rhinotomy and subtotal septectomy. (figure 3a and b) The patient was discharged to home the same day of surgery. The surgical specimen (figure 4a) was consistent with adenoid cystic carcinoma without bony or cartilaginous invasion and revealed clear margins. She did well postoperatively with minimal morbidity. She completed a postoperative radiation regimen of 8000 cGy over six weeks to the operative bed. At her last visit, 8 months post op, she had no evidence of recurrence on repeat imaging.

DISCUSSION

• Cancers arising in the sinonasal cavity and the base of skull are extremely rare, they comprise approximately 3% of all aerodigestive tract malignancies and less than 1% of all cancers. The incidence of malignant tumors of the nasal septum is especially low, with estimates of 2.7-8.4% of nasal and paranasal malignant tumors.1

• Early manifestations often masquerade as benign diseases such as rhinitis and sinusitis, due to the common complaints of nasal obstruction and recurrent infections. The anatomic location often allows for a delay in diagnosis due to the relatively unopposed growth in the large air filled spaces of the sinuses. In addition, this tumor can easily be mistaken for a sepal deviation given its indolent nature and its smooth, bulging appearance.

• Given its close proximity to critical structures, such as the skull base, dura, brain, carotid arteries, cranial nerves and orbits, treatment of locally advanced disease can have profound effects.

• Only 3 described cases have been reported in the English literature. With the overall paucity of data in the literature, treatment strategies are lacking. We describe a case of a 70 year old female with ACC of the nasal septum and review pathologic findings and treatment considerations.

• Historical findings can help to prognosticate local recurrence and overall survival. Complete surgical excision is the preferred method of treatment, but typically adjuvant radiation therapy is delivered.

• ACC of the nasal septum is an especially rare malignancy.

• Most common presenting symptoms include epistaxis and nasal obstruction, but atypical nasal symptoms should also be assessed.

• Similar to ACC of other sub-sites, there is a propensity to perineural invasion and metastatic sites typically occur to the lung.

• Histologic findings can help to prognosticate local recurrence and overall survival.

• Complete surgical excision is the preferred method of treatment, but typically adjuvant radiation therapy is delivered.