Parotid Oncocytoma Causing Newborn Airway Obstruction

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I Abstract

Objective: 1. Report the first published case of neonatal airway obstruction due to a congenital cystic parotid oncocytoma. 2. Describe the histologic features of salivary oncocytic neoplasms and propose a new etiology for tumorigenesis.

Methods: A single case at a tertiary care hospital is reported. A 6-week-old male presented with episodic airway obstruction and stridor. Direct laryngoscopy revealed medial compression of the lateral pharyngeal wall with significant airway compromise. Imaging demonstrated a cystic mass deep to the parotid, extending into the parapharyngeal space. Surgical method and outcome is discussed, including a histopathologic analysis of salivary oncocytosis.

Results: Oncocytic neoplasms are rare salivary gland tumors most commonly seen within the parotid gland. Oncocytoma is the most frequent morphology. The etiology of oncocytic neoplasms is obscure, although tumorigenesis may be secondary to both metaplasia and acquired mitochondrial dysfunction. We present the first known published case of a cystic parotid oncocytoma with airway obstruction in an infant. Imaging was crucial for preoperative planning. Surgical excision with facial nerve dissection was curative.

Conclusions: This case demonstrates that salivary oncocytic neoplasms can be present and symptomatic at birth, suggesting that congenital molecular mechanisms rather than acquired mitochondrialopathy may play a role in tumorigenesis.

II Background

- Oncocytes are epithelial cells characterized by abundant eosinophilic, granular cytoplasm filled with multiple tightly packed mitochondria showing varied pleomorphism
- Oncocytic neoplasia of the parotid is a rare form of salivary gland disease with an uncertain etiology, almost exclusively described in adults
- World Health Organization histologic classification of parotid tumors recognizes three types of oncocytic neoplasia
  - Oncocytoma is the most common morphology, followed by oncocytosis and oncocytic carcinoma
- Mitochondriopathy, either secondary to cellular nuclear DNA defects or actual mitochondrial DNA mutations, is thought to play a role in the development of oncocytic neoplasia
- Functional mitochondrial exhaustion and acquired dysfunction then occurs, but the exact molecular mechanism remains to be described
- Although acquired mitochondrialopathy is a plausible etiology in adults, this does not account for newborn oncocytopathy

III Objective

- To report the first published case of neonatal airway obstruction due to a congenital cystic parotid oncocytoma.
- To describe the histologic features of salivary oncocytic neoplasms and propose a new etiology for tumorigenesis.

IV Methods

- A single case at a tertiary care hospital is reported. A 6-week-old male presented with progressive airway obstruction. Imaging demonstrated a large parapharyngeal cyst, extending to the skull base and crossing the midline. This was excised using a retromandibular transparotid approach.

V Results

- Surgical excision was curative. Full facial nerve function was preserved. The mass was excised leaving the majority of the parotid gland intact. Histology confirmed a cystic parotid oncocytoma. Recent follow-up has revealed no recurrence.

VI Case Report

- Seven week old male presented to clinic with a three week history of nocturnal hypopnea, snoring and questionable apnea. No history of fever, infection, weight loss, cyanosis, or failure to thrive. 
- Past medical history for the patient and his mother was otherwise negative.
- No family history of tumor or craniofacial syndrome.
- Physical examination did not reveal a neck mass, but there was fullness below the left mandible. Flexible endoscopy demonstrated a normal epiglottis and vocal fold motion was intact. Stertor was noted, but no stridor.
- Direct laryngoscopy revealed medial displacement of the left lateral pharyngeal wall and soft palate. Larynx and subglottis were normal.
- The patient was kept intubated for an immediate neck CT scan.
- Based upon the CT findings, the patient was scheduled for transcervical / transparotid excision of the left parapharyngeal space mass.
- CT: Large cystic lesion noted which displaces the lateral wall of the nasopharynx medially and exhibits significant mass-effect on other surrounding structures. This extends from the skull base into the nasopharynx and subcutaneous tissues, occupying the superior aspect of the parapharyngeal space. There is medial displacement of the lateral pharyngeal wall with airway compression. There is no enhancement or inflammatory change surrounding the lesion to suggest an abscess. There is posterior displacement of the internal carotid artery.

VII Imaging

- Benign cyst that is partially lined by bland cuboidal to columnar epithelial cells occasionally forming papillary tufts. Oncocytes are seen with granular cytoplasm and abundant mitochondria.
- In other areas the lining is fibrous walled or endothelial lined.

VIII Pathology

- Benign cyst that is partially lined by bland cuboidal to columnar epithelial cells occasionally forming papillary tufts. Oncocytes are seen with granular cytoplasm and abundant mitochondria.
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IX Intraoperative Findings

- Cystic mass originating from the parotid (green arrow) with main trunk of the facial nerve (yellow arrow) draped over the mass
- Defect after mass removal shows extension into parapharyngeal space and a thin layer of remaining intact pharyngeal mucosa

X Conclusion

- Newborns do not have the environmental exposure history to acquire DNA defects or lifespan to acquire functional mitochondrial exhaustion or oxidative damage.
- Congenital, not acquired, mitochondriopathy is a plausible mechanism to explain parotid oncocytoma formation in a newborn.