Pediatric Sinonasal Chondromesenchymal Hamartomas: Case Report and Operative Considerations for Craniofacial Growth Optimization in Expanded Endonasal Approaches

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Objectives

- Review chondromesenchymal hamartomas (NCMH) in pediatric patients
- Examine the effect of expanded endoscopic endonasal surgeries on craniofacial growth, with the objective of understanding which structures are critical to preserve during surgery to minimize surgical morbidity while achieving maximal tumor resection.

Introduction

Nasal chondromesenchymal hamartomas (NCMH) are rare, benign sinonasal lesions treated with complete surgical resection.[1] For pediatric patients with NCMH or other skull-based sinonasal tumors, it is important to preserve structures critical for craniofacial development as the cranial vault continues to undergo changes over the first decade of life. Here we present a case report of a pediatric patient with NCMH and perform a literature review on surgical considerations for preserving structures critical for normal craniofacial growth.

Case Description

3-year-old female with PMH of iron deficiency anemia presented with several days of snoring, enlarged right nose, right eye proptosis, blood-streaked mucus, and low appetite. The patient underwent an outpatient tonsillectomy and adenoidectomy which was aborted due to finding of a protruding right nasal mass which was biopsied with indeterminate results. She was admitted for CT face and MRI face/brain with contrast (Fig. 1), with finding of large right sinonasal mass with possible involvement of anterior skull base without evident transdural extension. ENT, Ophthalmology, Hematology/Oncology and Neurosurgery were consulted. Patient underwent expanded endoscopic endonasal approach (EEA) for resection of mass without complications.

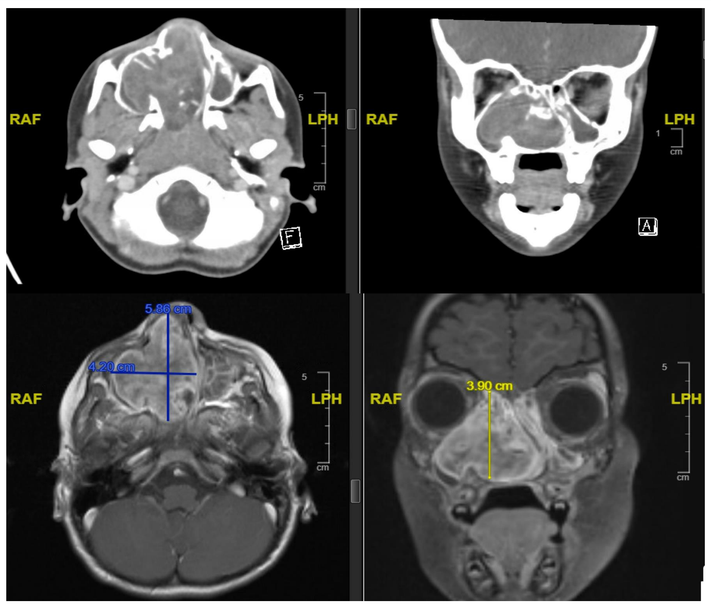

Figure 1. CT face with IV contrast and MRI T1 sequences with contrast showing large heterogeneously enhancing right nasal cavity mass extending into maxillary sinus and compressing right orbit. Mass was abutting the anterior skull base with possible bony involvement of the skull base but without evident intracranial transdural extension.

Outcomes

- Post-operative MRI (Fig. 2a) showed gross total resection of sinonasal mass.
- Final pathology report from surgery consistent with chondromesenchymal hamartoma.
- Based on final diagnosis, patient required no further oncological treatment apart from genetic testing; negative for DICER1 mutations.
- Based on 6-month surveillance MRI brain results suggestive of peripheral enhancement of prior tumor cavity (Fig. 2b), patient underwent endoscopic re-resection with margins negative for residual tumor.

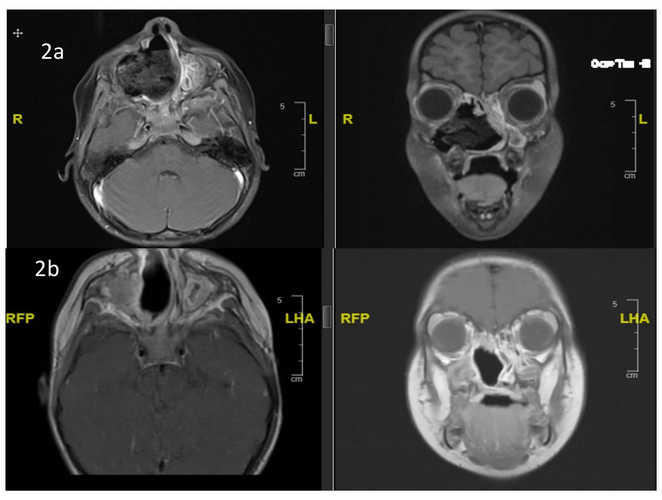

Figure 2. a) Postoperative MRI brain with contrast. b) 6-month surveillance MRI brain with contrast showing peripheral enhancement of right maxillary sinus and superior enhancement of right nasal cavity.

Sinonasal Chondromesenchymal Hamartomas

- Benign, locally destructive sinonasal tumor[1]
- Symptoms: nasal congestion, nasal obstruction, epistaxis, pain, and/or ocular symptoms (e.g. exophthalmos, epiphora) (Fig. 3)
- Differential diagnosis includes rhabdomyosarcoma, lymphoma, chondrosarcoma, meningoencephalocele
- Epidemiology: rare (~52 reported cases), infants / young children, M>F
- Etiology: genetic predisposition (DICER1 mutation)
- Diagnosis: Based on histopathology
- Pathology: Hyaline cartilage lobules in stromal background, S100+[2,3] (Fig. 4)
- Treatment: Complete resection, to prevent recurrence

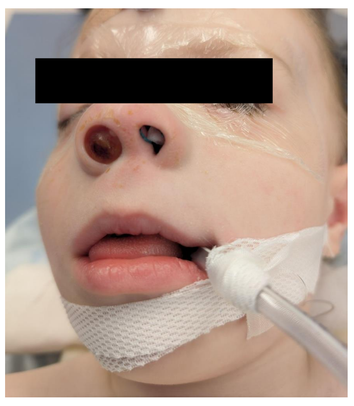

Figure 3. Pre-operative picture of mass protruding from right nasal cavity

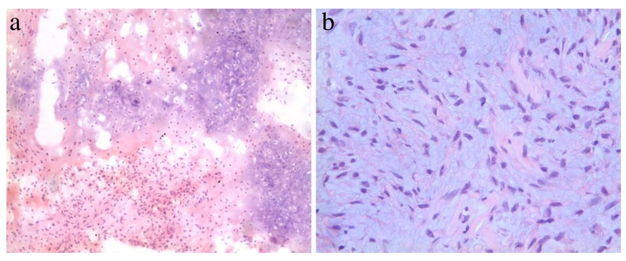

Figure 4. Example of NCMH histopathology with hyaline cartilage merged with spindle cells (A) and spindle cells loosely arranged in myxoid stroma (B) (excerpt from Wang et al. 2022)[2]

Special Pediatric EEA Surgical Considerations

- Neonates are solely nasal breathers until ~6 weeks of age.
- Lower tolerance for intraoperative blood loss.
- Smaller nasal aperture can limit access.
- Lack of sphenoid sinus pneumatization can require extensive drilling during approach and increase surgical risk due to lack of anatomic landmarks.[3,4]
- Inter-carotid distances at the cavernous sinus and clivus can be narrow and variable, potentially increasing operative risk.[5]
- Higher risk of cerebrospinal fluid (CSF) leaks compared to adults due to reconstructive challenges and anatomic variability.[6]

Preserving Craniofacial Growth in EEA Surgery

1. Identification and preservation of facial buttresses → Despite resection of tumor involving nasolacrimal duct, preserved medial maxillary buttress
   - Medial maxillary / nasomaxillary pterygoid buttress: vertical buttress which dictates midfacial structure growth (Fig. 5).
   - Disruption of medial maxillary buttress can cause midfacial deformity, via loss of lateral alar support and nasal ala retraction. Avoid endoscopic Denker approach for intratemporal and pterygopalatine fossa)[7,8]
2. Orbit preservation → During tumor resection, preserved inferior orbit periosteum / inferior-medial orbital strut
   - Preservation of inferior periorbita to prevent inferomedial displacement of globe position, preserve orbital height, decrease enophthalmos risk
3. Maximize nasal septal preservation if possible: avoid incisions through growing/supporting zones, i.e. sphenodorsal zone (for height/length of nasal bones) and sphenospinal zone (for forward growth of maxilla)[9]

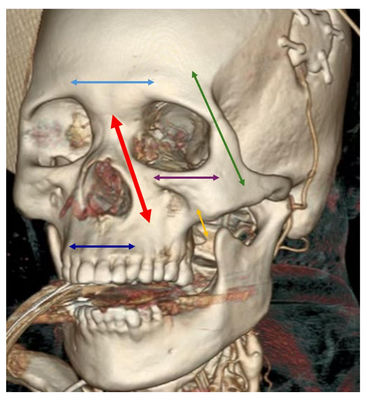

Figure 5. 3D CT image of adult facial skeleton depicting facial buttresses (based on reference image from Reiter et al. 2017). Vertical buttresses are medial maxillary / nasomaxillary (red), zygomaticomaxillary / lateral maxillary (green), pterygomaxillary / posterior maxillary (yellow) buttresses. Horizontal buttresses are superior orbital rim (light blue), infraorbital / upper transverse maxillary (purple), maxillary alveolar / lower transverse maxillary (blue) buttresses.

Conclusion

During expanded endonasal surgical approaches in the pediatric population for skull base tumors such as NCMH, knowledge of anatomical landmarks and the key structures involved in craniofacial development is necessary to prevent midfacial deformities and achieve the goal of complete tumor resection.

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