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Soft palate teratoma: 5 month old presenting with failure to thrive and severe obstructive sleep apnea

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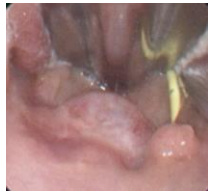
Introduction

Oropharyngeal teratomas are an extremely rare congenital tumor. They are often diagnosed prenatally and can cause significant airway obstruction and feeding difficulties at birth. We present a 5-month-old female that was diagnosed with a palatal teratoma that presented with failure to thrive, difficulty feeding and eventually with severe obstructive sleep apnea.

Timeline

- Patient was born with a normal birth weight. She breastfed successfully. Mother reports stridor since birth, worse with feeding.
- After an episode of RSV, she continued to have feeding difficulties with increased work of breathing during feeds. This resulted in failure to thrive.
- She was seen in consultation with Lactation specialist who noted retrognathia and poor tongue protrusion. Referred to ENT.
- ENT performed flexible laryngoscopy noting laryngomalacia
- An NGT was placed to assist with feeding with mom continuing to breastfeed. There was initial weight gain, but vomiting led to further weight loss. NGT removed 6 weeks later with no significant weight gain.
- She presented to CMH with severe stridor, failure to thrive, and was in the 0.07th percentile for weight .
  - → presented to CMH Kansas, initial scope revealed no abnormalities
  - → transferred to CMH Main, bilateral nasal endoscopy showed palatal mass obstructing left oropharynx
  - →Polysomnogram reported AHI of 172.5 and oxygen nadir of 74%
  - → MRI revealed a 2.2 x 2.2 x 1.8cm expansile left soft palate mass with significant extension into the soft palate musculature



- Taken to the operating room for surgical debulking of airway and biopsy which resulted as teratoma with glial and intestinal tissue present.
- One month later taken to OR for complete resection of remaining teratoma tissue with primary closure of palate. She is scheduled for initial post-operative MRI in 3 months. Patient following closely with Otolaryngology and hematology/oncology.

Investigation

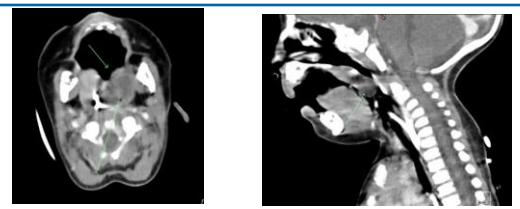


Figure 1: CT neck with contrast soft tissue

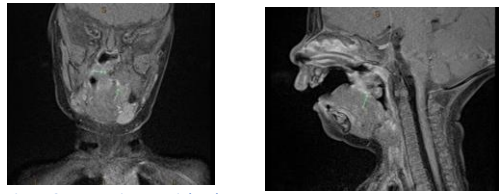


Figure 2: MRI Brain T1-weighted post contrast

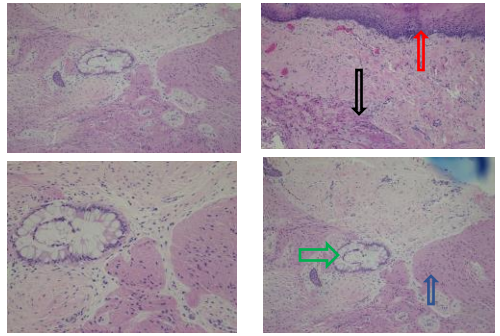


Figure 3: Surgical pathology slides (red: surface epithelium of palate, black: mature glial tissue, green: intestinal tissue, blue: mature glial tissue).

Discussion

Teratomas occur in 1:4,000 births and display a female predominance. The head and neck region only represents 5-15% of these tumors and only 2% are in the oropharynx.[6] There is no pathognomonic characteristics found on imaging, however calcifications can be suggestive of teratoma.[3] The diagnosis is made by pathologic identification of two of the three germ cell layers.[5] Identification of various tissues such as bone, muscle, exocrine glands, solid organs, intestinal tissue, neuroglial, skin, and teeth are possible within the tumor. Ectopic germ-cells undergo proliferation at the 4th or 5th week of gestation. They differentiate into mature or fetal tissue creating delineation between mature and immature teratoma.[3]

Although most teratomas are benign, there is potential for malignant transformation involving any of the represented germ cell layers. Many teratomas are diagnosed prenatally and can be quite large, often requiring Ex Utero Intrapartum Treatment or, EXIT, procedure at birth to establish a safe airway. Most current case reports in the literature discuss patients in the newborn timeframe presenting with airway obstruction or other concerning findings.[4] Complete surgical excision is considered gold standard regardless of the presentation age.[1]

Conclusion

Overall, this case highlights the importance of a thorough head and neck exam including a bilateral flexible laryngoscopy when evaluating an infant with airway obstruction. Providers evaluating these patients should consider oropharyngeal masses, such as teratoma as part of the differential to ensure accurate and timely diagnosis.

References

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