Oropharyngeal Dermoid Cyst in an Infant with Intermittent Airway Obstruction

A Case Report

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SUMMARY – *Dermoid cysts are benign epithelial inclusions and cystic lesions that may occur in several locations including the oropharynx. We describe the case of a two-month-old baby girl who presented with progressive respiratory distress, hypoxemia, and feeding difficulties because of an oropharyngeal dermoid cyst. The child had an airway work-up that included laryngoscopy. However, the mass remained undetected. This is most likely explained by the mobile nature of the lesion, prolapsing into the high nasopharynx in supine position. In our patient, magnetic resonance imaging (MRI), initially performed to rule out brainstem pathology, revealed an oropharyngeal dermoid cyst. This case shows the potential role of neuroimaging in the diagnostic work-up of a young child presenting with respiratory distress by excluding a central nervous system lesion and diagnosing an “unexpected” nasopharyngeal mass lesion. In addition, MRI allowed exclusion of skull base lesions of neural origin such as an anterior meningoencephalocele or heterotopic neuroglial tissue which would be managed differently from pharyngeal masses.*

Introduction

Infants with upper airway obstruction at the pharyngeal level often present with inspiratory stridor or stertor and feeding difficulties. Masses are rare causes of upper airway obstruction and have to be suspected within the entire upper airway including naso- and oropharynx, larynx, and trachea. The differential diagnosis includes a variety of rare lesions such as lymphangioma, hemangioma, glioma, encephalocele, meningocele, teratoma, dermoid cyst, branchial cleft anomaly, neurofibroma, sarcoma, craniopharyngioma, and neuroglial heterotopia¹.

We describe a two-month-old infant who presented with intermittent airway obstruction and feeding difficulties due to an oropharyngeal dermoid cyst and discuss the key role of head MRI in making the diagnosis.

Case Report

A two-month-old female infant presented to the emergency department (ED) of our tertiary Children's Hospital with worsening respiratory distress and hypoxemia. She was born at term by Caesarean section because of severe oligohydramnios and breech presentation. On delivery she was floppy, apneic, and bradycardic (heart rate of 60/min). Micrognathia was noted. She was intubated within the first minutes of life for persistent apnea and bradycardia, but she was extubated electively when she stabilized within the first hour of life. At day of life (DOL) 4, she developed mild inspiratory noise during sleep and agitation. Nasal edema was suspected, a probatory treatment with prednisolone nasal drops was initiated, but showed only minimal benefit. By DOL 11, the respiratory distress increased with worsening subcostal retractions

Figure 1 A) Parasagittal T1-weighted image shows a mixed hypo- and hyperintense oropharyngeal lesion (arrows). B) Axial T2-weighted image shows hyperintense signal of the same ovoid lesion (arrows). C) Intraoperative image shows forceps catching the peduncle of the mass lesion. D) Macrophathology specimen of the mass lesion shown to be a dermoid cyst.

and intermittent oxygen desaturation episodes to about 85%. Fiberoptic laryngoscopy was performed and showed patent nasal cavities, no choanal atresia, unremarkable naso- and oropharynx, and normal vocal cord anatomy and motion. Her respiratory status stabilized and feeding improved, so she was discharged home at DOL 22.

At two months of age, the baby girl presented again to the ED with respiratory distress. According to her parents she had had

no respiratory noise since discharge before this presentation, but did notice mild tachypnea during feeds. Three days prior to admission the tachypnea increased and episodes of non-bilious vomiting occurred. In the ED, the patient had persistent hypoxemia on room air with oxygen desaturation to 85% SO_2 . A supplemental oxygen support via a four-liter oxygen high-flow nasal cannula was started, and she was admitted to the pediatric intensive care unit for further work-up and therapy.

The further work-up included echocardiography and suspension microlaryngoscopy and rigid bronchoscopy performed under general anesthesia with a 5 cm Parson's laryngoscope which showed normal oropharyngeal, laryngeal, and distal airway anatomy. Additionally, naso-duodenal feeding was started to rule out potential microaspirations, but her respiratory status remained tenuous with noisy breathing and the need for high-flow oxygen support. Brain MRI was performed to investigate possible intracranial causes of airway disease, specifically brainstem abnormalities. This study showed a $1.7 \times 1.5 \times 2.4$ cm ovoid lesion located dorsal to the uvula and separated from the adjacent skull base. The lesion showed a mixed hypo- and hyperintense signal on T1- and a hyperintense signal on T2-weighted as well as on fluid attenuated inversion recovery (FLAIR) images (Figure 1A,B). No restricted diffusion was seen on diffusion-weighted imaging. The brainstem was normal, and the pharyngeal mass had no intracranial tracts. Further inspection of the oral cavity revealed a pharyngeal mass, suspected to be a dermoid cyst or teratoma, filling a large portion of the posterior oropharynx. This was defined additionally in the operating room the following day, where it was found to be attached to the soft palate margin laterally. It was excised without difficulties (Figure 1C,D). Pathology confirmed the suspicion of a dermoid cyst. The baby girl was discharged home soon after surgery with normal respiratory function.

Discussion

Dermoid cysts are benign epithelial inclusion cystic lesions thought to be formed in the third to fifth gestational week as the result of abnormal separation of ectoderm and neuroectoderm². Dermoid cysts typically contain dermal skin appendages such as hair follicles, sweat glands, and, rarely, teeth, and grow by both desquamation and glandular secretions^{2,3}. Dermoid cysts may occur in several locations including almost everywhere on the skin (particularly around the eyes), in the abdominal cavity and gonads, everywhere in the oropharyngeal cavity (particularly on the floor of the mouth), in the head and neck regions, in the spinal canal, and intracranially (most frequently in the posterior fossa, either within the vermis or in the fourth ventricle)^{4,5}.

Imaging is helpful to localize the cyst and

assess its origin and extent and is essential to characterize the lesion, determine a potential intracranial extension, and differentiate it from other lesions that may occur in the same location. On computed tomography, dermoid cysts appear as fat-density masses that do not show post-contrast enhancement unless they are infected. On MRI, dermoid cysts have typically a hyperintense signal on T1- and T2-weighted as well as on FLAIR images. The MR signal is typically suppressed after a fat suppression pulse is applied. In contrast to epidermoid cysts, dermoid cysts typically do not show restricted diffusion.

Dermoid cysts in the oropharyngeal cavities may present with respiratory distress and feeding difficulties as in our patient⁶. Other symptoms include hearing loss, otorrhea, vomiting, earache, bleeding, snoring, and recurrent ear infections⁶.

Small lesions may remain asymptomatic. Oropharyngeal dermoid cysts are typically diagnosed by direct or indirect visualization. In the case of a high clinical suspicion of oropharyngeal mass lesions and negative laryngoscopy, head and neck imaging is a helpful diagnostic tool as shown by our case.

Neuroimaging allows the differentiation of dermoid cysts from other lesions within the oropharynx such as meningoencephalocele, heterotopic neuroglial tissue, glioma, and craniopharyngioma (Figure 2)^{1,6}. This differentiation is essential in terms of surgical management and outcome. In particular, an intracranial extension should be evaluated. Meningoencephaloceles show a direct communication through a skull base defect with the cranial vault and have a hypointense signal on T1-weighted and FLAIR images, similar to brain tissue. The most common location of skull base meningoencephaloceles is frontoethmoidal (80%), while sphenoethmoidal meningoencephaloceles mimicking an oropharyngeal dermoid cyst are rare (1-2%)⁷. Heterotopic neuroglial tissue is a rare malformation composed of differentiated derivatives of neuroectodermal tissue that include disorganized neuronal and glial cells¹. Heterotopic neuroglial tissue may be located within or, most commonly, outside the cranial vault. It is most frequently found in the nasal cavity, but it has also been reported in the orbit, pterygopalatine fossa, middle ear, neck, thorax, and pharynx¹. Heterotopic neuroglial tissue has a typical neuroimaging appearance with intermingled T1 and T2 signal characteristics of brain gray and white matter with small

Figure 2 A) Parasagittal T1-weighted image shows a hypointense encephalocele extending through the floor of the sella turcica into the superior aspect of the posterior pharynx (arrows). B) Axial T2-weighted image shows hyperintense signal of the encephalocele (arrows). C) Coronal T2-weighted fast spin-echo (FSE) images. Heterotopic brain tissue presents as an inhomogeneous, partially T2-hyper-hypointense lesion within the right parapharyngeal space. Arrows: hypointense, solid components extend along the carotid artery and the maxillary nerve into the right cavernous sinus. D) Axial T2-weighted images. The left parapharyngeal component is moderately hyperintense (arrows) and displaces the right parotid gland laterally and the pterygoid muscles anteriorly. The parapharyngeal component is partially isointense to the brain parenchyma. C and D are reprinted with permission from Huisman TA¹.

enclosed cysts. Nasal gliomas are thought to represent accumulations of dysplastic brain tissue in the nasal cavity or subcutaneous tissue and are separate from intracranial tissue⁸. Hypointense signal on T1-weighted images usually differentiates between nasal gliomas and oropharyngeal dermoid cysts.

In conclusion, our case shows how head and neck MRI may be helpful in the diagnosis of

suspected oropharyngeal mass lesions and in the differential diagnosis of children with respiratory distress secondary to a variety of lesions within the region of the skull base or pharynx. The differentiation between dermoid cysts and other lesions such as an anterior meningoencephalocele or heterotopic neuroglial tissue is essential in terms of management and outcome.

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