

Epidermoid Cyst of Uvula: A Rare Cause of Upper Airway Obstruction

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Abstract

A three months old female baby presented to the emergency with complaints of excessive crying, rapid breathing and high - grade fever for one day. On arrival, the patient had respiratory distress with endotracheal tube in situ and bilateral crepitations on auscultation. Oral cavity examination revealed a well-circumscribed, white colored mass with a smooth surface extending from the uvula. The baby was managed successfully by surgery and the baby has been asymptomatic on follow up.

Introduction

Abnormal epithelial components of ectodermal tissue lead to formation of epidermoid cysts, which usually develop in antenatal lifetime. In some cases they may be acquired after surgery or trauma.¹ only around 2% occurs in the oral cavity, and the most common site being the floor of mouth. There occurrence in the soft palate and uvula is rare.² Although most are asymptomatic and are found by chance, those in oral cavity may present with symptoms in the neonatal and infancy period.³ We present here a rare case of an epidermoid cyst of uvula presenting with feeding and breathing difficulty in early infancy

Case report

A three-months-old female baby with uneventful antenatal and natal history presented to the emergency with complaints of excessive crying, rapid breathing and high-grade fever for one day. The baby had dry cough from past two days which increased during feeding. Parents sought a medical opinion for the same near their home, and the patient was intubated by a local physician just prior to admission in our hospital. On arrival in our hospital, the patient had respiratory distress with a rate of 84 per minute, with endotracheal tube in situ and bilateral crepitations on auscultation. Heart rate was 160 per minute (with no other abnormal cardiovascular findings), with normal capillary refill time. Patient was put on ventilator support in view of suspected severe pneumonia and started on intravenous antibiotics and fluids. All necessary investigations were sent complete blood count report showed increased total leucocyte count 25980 cells

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/ mm³ with neutrophilic predominance, C-Reactive Protein was negative, Chest X-ray was suggestive of aspiration pneumonitis right side. Patient improved gradually and was extubated after 48 hours of admission. Feeds were started by orogastric tube initially and shifted to spoon feeds after extubation. However, significant feeding difficulty was noted once feeding by spoon was initiated, and her breathing was observed to be labored when she was put in supine position [Figure 1]. History was again reviewed from mother and she reported that patient had suffered repeated episodes of cough during feeding (since one month age), along with breathing difficulty when put in lying down position. This made us suspect strongly of some upper respiratory tract abnormality and the patient was evaluated for same with the help of the otolaryngorhinology team. Oral cavity examination revealed a well-circumscribed, white colored mass with a smooth surface extending from the uvula [Figure 1]. A non-contrast computed tomography (CT) of the region was performed which revealed a well-defined, smooth-margined, round to ovoid, fat-attenuating, midline lesion measuring up to 45 mm X 30 mm X 25 mm arising from the soft palate replacing the normal uvula. No evidence of any internal air or blood was noted within the above-described lesion. No other associated abnormality including hard palate erosion was noted. Based on the above findings, the diagnosis of uvular dermoid cyst was made [Figure 2].



Figure 2: Sagittal section of CT head and neck showing a mass arising from uvular region (arrow)

No other external facial or neck cysts, sinuses or lesions were noted and the rest of the intraoral space was unremarkable.

Patient was referred to paediatric surgeon for surgical management of the mass. The mass was removed successfully without any intra-operative and post-operative complications. The excised mass was sent for histopathological examination which showed normal skin with adnexal glands, dermis showing proliferation of mature adipose tissue with septa of fibrocollagenous tissue and striated muscle fibres, deeper tiny foci showing pseudostratified columnar epithelium with few normal salivary glands. Patient visited our hospital again after one week of surgery for follow-up and there were no signs of any complications. Patient no more had any difficulty in feeding and is currently doing fine on follow up.

Discussion

Epidermoid uvular cysts are benign developmental abnormalities which arise from abnormal epithelial components of ectodermal tissue during in utero period. Such cysts may be congenital or acquired during the lifetime.^{4,5} In early fetal life, between 6th and 12th weeks of gestation, buried epithelial cells during fusion of the first and second branchial arches, lead to formation of epidermoid cysts of the uvula and soft palate.⁶

Cysts can develop anywhere in the body; the overall incidence reported in the head and neck region is near 7.0%. In this region, the most common locations are the orbit (46.6%), submental and the floor of the mouth (23.3%), the nose (12.6%), the neck (10.7%), and the lips



Figure 1: A white smooth surface lined mass seemingly attached to soft palate region (on oral examination)

(2.9%).⁴ Uvular cysts usually don't develop any symptoms and are incidental findings. Some newborns and infants may present as difficulty in deglutination and sucking.⁷ Due to the closeness of the nasopharynx to the uvula, this has a potential to cause obstruction of airway.

Differential diagnostic possibilities of uvular cysts include, such as epidermoid cysts, mucoceles, lymphoepithelial cysts, minor salivary gland neoplasms, and cystic teratomas. Epithelial cysts have squamous epithelium as their lining in the absence of adnexal structures. Fine needle aspiration cytology is often not done in uvular masses because of their anatomical location and potential to cause increase of swelling. If non benign nature is suspected, then MRI is used for pre-operative approach of the mass. Imaging may not be required if the mass appears to be localised with fully appreciated borders on examination.⁵ Surgical approach is required for large mass causing problems in feeding or respiration.

Conclusions

Feeding difficulty and respiratory distress has many differentials in infancy but dermoid cyst in uvula is an uncommon cause of such clinical presentation. Any patient presenting with difficulty in breathing and feeding should undergo oral cavity and expert throat examination to rule out this rare entity.

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