LARGE DERMOID CYST OF THE TONGUE IN A NEONATE: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Background: Dermoid cysts of the tongue are very rare masses of the oral cavity derived from ectodermal elements. These tumours are benign and slow-growing, typically asymptomatic but large cysts that fill most of the oral cavity can cause complications of dyspnoea, dysphagia, respiratory obstruction, and difficult intubation. The definitive treatment is surgical excision. The aim of this report is to demonstrate that large intralingual cyst located at the anterior two-thirds of the tongue can present without difficulty in breathing, unlikely to have difficult intubation and, early excision can give a good outcome.

Patient: The patient is a new-born male who was brought to the Emergency Paediatric Unit of the hospital 13 hours after birth with a huge swelling in the tongue protruding outside the oral cavity. There was difficulty feeding, however no difficulty in breathing and no other congenital anomalies. The swelling obliterated most of the anterior two-thirds of the tongue up to the foramen caecum. Ultrasound and magnetic resonance imaging were diagnostic of dermoid cyst.

Intervention: The patient had surgical excision of the cyst under general anaesthesia on the 15th day of life. Histology was in keeping with a lingual dermoid cyst .

Conclusion: This is a rare large intralingual cyst successfully managed by early excision with an uneventful post-operative course.

Key words: Neonate, Tongue, Large congenital dermoid cyst, Surgical excision

Introduction

Dermoid cysts are benign congenital lesions of ectodermal origin.[1] They are caused by the encasement of ectoderm at the time of closure of an embryogenic tissue.[2] They contain sebaceous glands and secretions, and rarely ,may contain hair and hair-like follicles.[2] Dermoid cysts contain hair follicles and sebaceous glands but epidermoid cysts are lined by epithelium, while teratoma contains mesodermal elements.[3] Congenital cystic lesions are commonly found throughout the body but are rare in the floor of the oral cavity.[1-5] The majority of reported cases are in the midline of the body especially in the testis and ovaries.[4] In the head and neck region they occur most frequently in the periorbital area with 6.5% of dermoid cysts involving the oral cavity.[4] Most oral dermoid cysts are located on the floor of the mouth and involvement of the tongue is rare.[6-9]

Dermoid cysts of the tongue are benign and slow-growing and may be asymptomatic but large cysts can cause complications such as inflammation or dysphagia, dystonia, and dyspnoea due to pressure effects.[5] Large cysts may fill the entire oral cavity and make tracheal intubation extremely difficult.[10]

Lesions that present as cyst in the floor of the mouth include neoplasms, infections and developmental processes i.e. lymphangioma, acute infection, neurofibroma, haemangioma, sublingual ranula, and lipomas.[6,11] Although rare, dermoid cyst should be included in the differential diagnosis of cystic lesions of the tongue.[6] The definitive treatment of these lesions is complete surgical excision[1,11] with a very low recurrence rate, and it is facilitated by the fibrous capsule surrounding the cyst that makes it easy to be enucleated.[11] The index patient presented with large intralingual cyst located at the anterior two-thirds of the tongue without difficulty in breathing had uneventful intubation and successful excision with good outcome..

Case Report

A 13-hour-old male neonate, born to a 38-year-old woman who was brought to the Emergency Paediatric Unit of the hospital with a large tongue noticed at birth. He was unable to close the mouth with difficulty in feeding. However, no difficulty in breathing or stridor and no associated anomalies on other parts of the body .The pregnancy was booked at a primary health care facility, had several uneventful visits, and the delivery was at term via spontaneous vaginal delivery.

Examination findings were those of a neonate with stable vital signs, Weight of 4 kg, Length of 60 cm and occipito-frontal circumference (OFC) of 36 cm. There was a large swelling within the tongue, from about the junction of the anterior two-thirds and posterior one-third to the tip of the tongue obliterating most of the oral cavity and protruding outside the mouth. It measured about 8 x 8 cm with healthy overlying mucosa. The swelling was soft, fluctuant, non-reducible, and non-pulsatile. (Fig 1) There was no other congenital anomalies.

Figure 1 .The large protruding tongue with lesion at birth

The following differentials were entertained; congenital haemangioma, ranula, lymphatic malformation, harmatoma, salivary gland tumour, and congenital dermoid cyst. The patient was co-managed with paediatricians in the special care baby unit of the hospital.

Ultrasound Scan revealed a cystic an-echoic swelling that was occupying most of the tongue, with multiple septations and tiny internal echoes that did not change colour on Doppler scanning. The mass measured 4.3 x 3.1 x 3.8 cm with a volume of 25.3 mls. Magnetic Resonance Imaging (MRI) findings showed a large well-defined, thin-walled lesion noted at the floor of the mouth and occupy most of the oral cavity with protrusion of the anterior aspect of the mass outside the oral cavity. It appears homogenously hypotense on T1 and fluid attenuated inversion recovery (FLAIR), and hyperintense on T2, measuring 4.5 x 5.0 x 6.2 cm in craniocaudal, transverse and anteroposterior dimensions respectively. It was observed to stretch and displace the genioglossus muscle upwards and backwards, and severely compress on the surrounding soft tissues. (Fig 2)

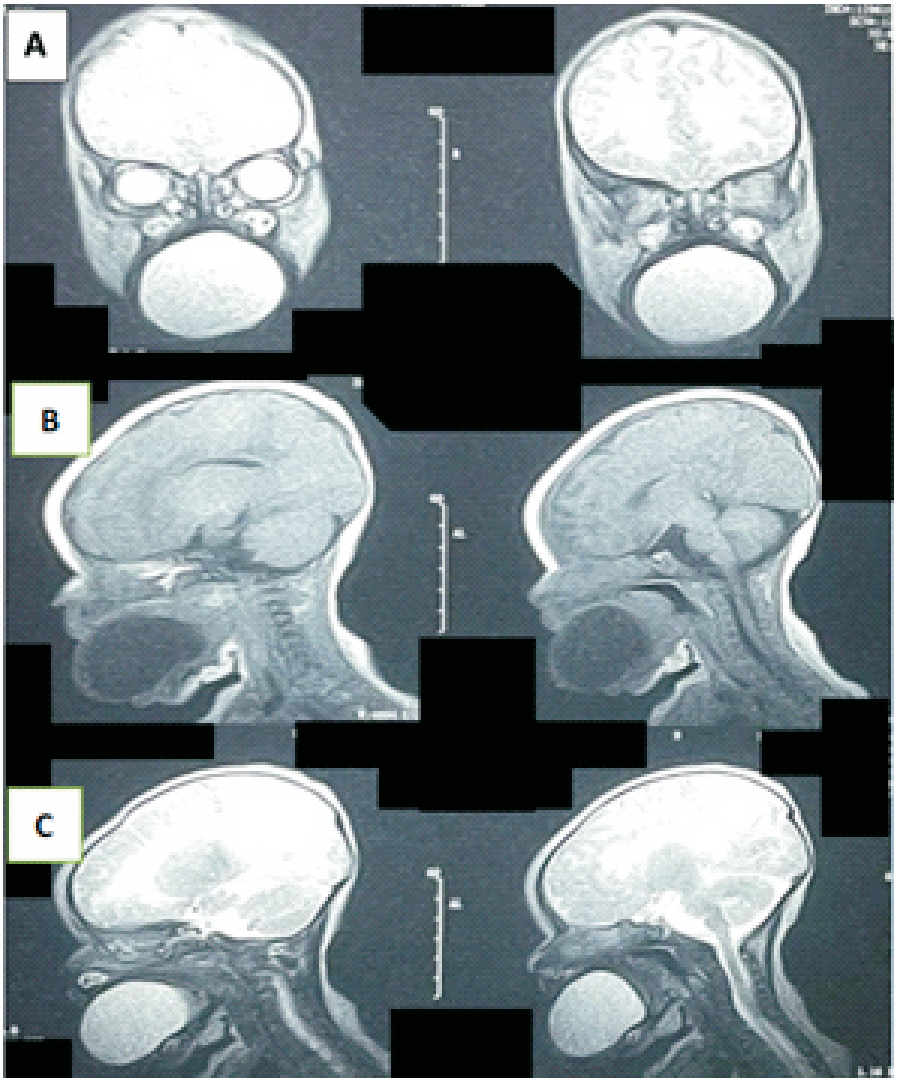

Figure 2; The cyst on MRI; Coronal T1W image showing a homogenously hypointense lesion(A), Sagittal T1W image showing a homogenously hypointense lesion(B) and Sagittal T2W image showing hyperintense lesion in keeping with dermoid of the tongue©

A diagnosis of a large congenital dermoid cyst of the tongue was made. Patient was prepared for and had surgical excision of the cyst on the 15th day of life, under general anaesthesia with successful endotracheal intubation. The excision was effected via a midline longitudinal incision on the ventral surface of the tongue. The submucosal tissue was bluntly dissected until the cyst wall was seen. The cyst was dissected from the tongue muscle and enucleated while safeguarding the lingual nerve. The cyst was a well-circumscribed, thin-walled sac with clear gelatinous content. The wound was closed in layers with absorbable sutures. (Fig 3a to 3c)

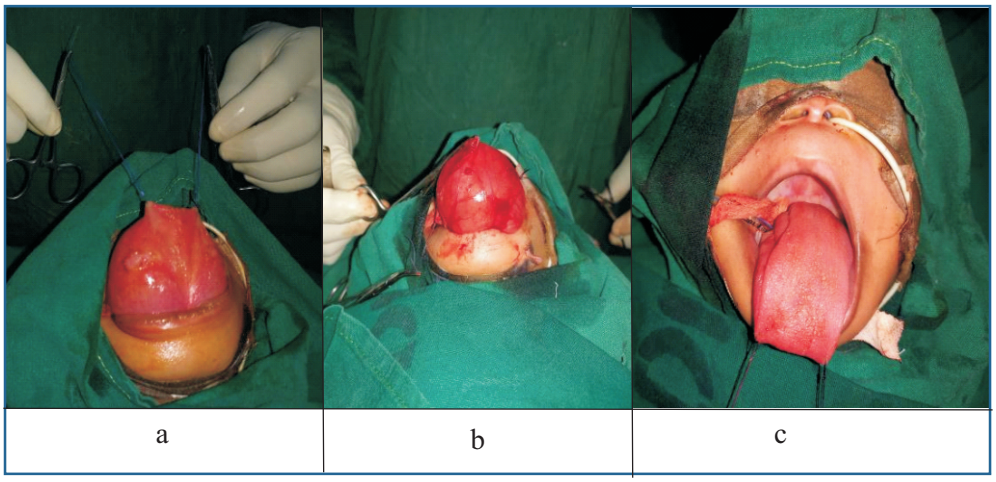

Figure 3: Sutures used to pull the tongue out of the oral cavity(a), Cyst demonstrated after blunt dissection(b), Tongue after closure of the cavity in layers(c)

He had uneventful post operative period and was discharged on day eight of surgery (24th day of life) with follow up visits at 2 weeks and 8 weeks of the discharge. (Fig 4)
The histology revealed features consistent with dermoid cyst.

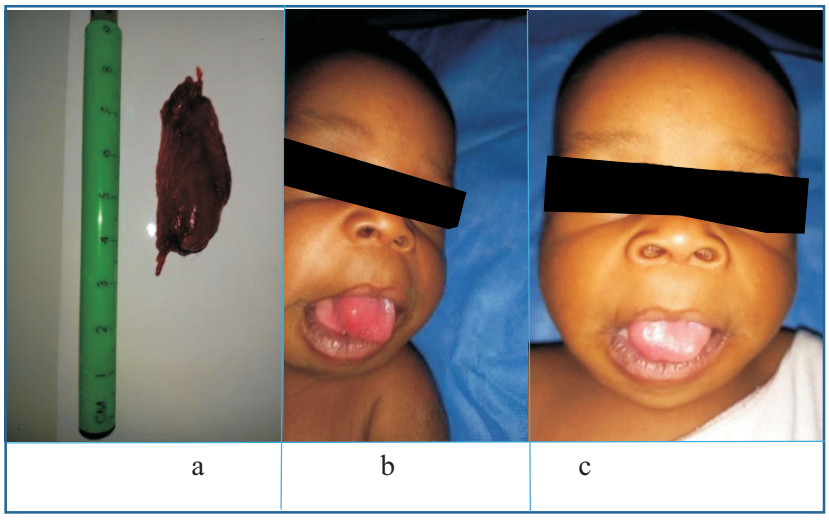

Figure 4: excised cyst sac(a), the tongue at 2 weeks after discharge(b), the tongue at 8weeks after surgery(c)

Discussion

Dermoid cyst is a term used to describe congenital cystic lesions where adnexae such as sweat glands, sebaceous glands and hair follicles are present.[12] They are believed to arise as a result of entrapped pluripotent cells during embryogenesis.[13] Histologically, they are lined by epidermis and are extremely rare in the tongue.[13] Based on histology dermoid cysts in the floor of the mouth are classified by Meyer into dermoid, epidermoid, and teratoid/teratomata cysts.[4] Dermoid cysts have a wall of stratified squamous cornified epithelium and contain smegma and keratin scales as well as cutaneous appendages such as hair follicles, hairs, and sebaceous and sudoriparous glands. However, epidermoid cysts have a wall of stratified squamous cornified epithelium and contain smegma and keratin scales without cutaneous appendages. Lastly, teratoid/teratomata cysts have a wall of stratified squamous epithelium with or without cornification and contain smegma and keratin scales as well as elements of the middle blastoderm such as vascular formations, elements of muscle and bone, dental tissues, or even whole teeth.[4] In this patient, the cyst was lined by stratified squamous epithelium, with foci of sebaceous glands, and numerous thin-walled blood vessels, which was consistent with a dermoid cyst. About 34% of dermoid cysts are found in the head and neck, of which 6.5% are located at the floor of the mouth.[5] and cyst of the tongue represent less than 0.01% of all oral cavity cysts.[11] Oral sublingual dermoids can vary in size from a few mm to 12 cm in diameter.[14] They are slow in growth, most commonly diagnosed in the first to third decades of life, and are either acquired (85%) or congenital (15%), both of which are histologically identical[14] with no gender predilection.[15]

The patients with dermoid cyst usually present early in life with asymptomatic mass that is slow growing[1] however the cyst in this patient was large occupying almost the entire oral cavity which affected feeding. Despite the size of the lesion, there was no difficulty in breathing and nasogastric tube was passed successfully. This was most likely because of the location of the cyst at the anterior two-thirds of the tongue. Embryologically, the tongue consists of two separate parts; the anterior two-thirds and the posterior one-third.[6] The mucous membrane of the anterior two-thirds of the tongue develops from the first branchial arch. Specifically, three swellings of primitive mesenchyme; the midline tuberculum impar and bilateral lingual swellings, merge to form a single mass from which the anterior two-thirds of the tongue is formed. The mucosa of the posterior third of the tongue is formed from a large midline swelling of mesoderm of the second, third, and fourth branchial arches.[6] Therefore, entrapment of ectoderm resulting in formation of the huge cyst in the anterior two-thirds of the tongue will not obstruct the posterior oral cavity, this explains why there was no difficulty in breathing and nasogastric tube was passed successfully.

Different imaging techniques have been reported as adjuncts in the diagnosis of dermoid cyst.[16,17] Histology remains the definitive diagnosis of dermoid cyst.[17] Ultrasound imaging is the initial diagnostic modality of choice for oral lesions.[5] Dermoid appear as well circumscribed, unilocular cysts that may contain either anechoic or hypoechoic regions or multiple echogenic nodules because of the presence of epithelial debris or skin appendages.[5] In this patient the Ultrasound revealed cystic anechoic collection that was occupying most of the tongue, with multiple septations and tiny internal echoes that had no colour changes on Doppler interrogation. Contrast enhanced Computer tomographic (CT) is a preferred method of imaging however MRI and plain CT allow more precise localization of the lesion and also enable the surgeon to choose the most appropriate approach.[16] Magnetic resonance imaging is an accurate modality for the diagnosis and follow-up of tongue dermoid cyst and has the advantage of having no radiation exposure, it can also delineate the cyst and demonstrate its extension throughout the floor of the mouth.[7]

The differential diagnosis of intralingual cyst include, but not limited to, the following; dermoid cyst, lymphatic malformation, haemangioma, thyroglossal duct cyst and bronchogenic cyst. *Dermoid cysts* are mainly seen in the midline of the body and in the testis and ovaries but the floor of the mouth is the commonest intraoral site and extremely rare in the tongue.[13] Histologically they are lined by epidermis and characteristically contain skin adnex.[13] Therefore, it should be considered as differential in congenital cystic swellings of the tongue.

The effective treatment is surgical excision/enucleation of the cyst under general anaesthesia[1,7,10,11,13,16,17] which could be via extraoral, intraoral, or both approaches depending on the anatomical location, and size of the lesion with the aim of complete removal and preventing recurrence.[3,11,17] However, giant extensive cysts abutting or involving vital structures might be considered for marsupialization.[3,6] Nevertheless, irrespective of the approach, special consideration must be given to the Wharton's duct, lingual nerve and vessels.[17] Though difficult intubation was anticipated, patient successfully had an endotracheal intubation which was uneventful. Oluleke et al.[1] who reported 14 cases of sublingual dermoid cysts experienced difficult intubation in most of the patients. Oftentimes, needle decompression of the fluid was done to reduce the volume of the cystic fluids to facilitate endotracheal intubation.[1,10] Postoperative management was uneventful except for mild tongue oedema without airway challenges which improved with dexamethazone for 24 hours. But Oluleke et al.[1] reported respiratory infection, respiratory obstruction, feeding difficulties and secretions in their series.

Conclusion

Giant lingual dermoid cysts in a newborn are rare. It can present with difficulty feeding, but without challenge in breathing and endotracheal intubation as in the index patient. However, when the cyst is located at the posterior aspect of the tongue or oral cavity, there may be considerable technical challenges to the anaesthesiologist and the surgeon.

Conflict of interest

The authors declare that there is no conflict of interest

Informed consent

The authors certify that the necessary and appropriate consent was obtained from the parents of the patient to publish the clinical information and images. The parents were made to understand that the name and initials will not be published and all efforts will be made to conceal his identity. However, anonymity will not be completely guaranteed.

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