

Case Report

Congenital Heterotopic Gastrointestinal Cyst of the Tongue in a New-born: Case Report

Oginni FO, Braimah RO, Oladejo T, Adenekan AT

Abstract

Heterotopic gastrointestinal cyst is an extremely rare clinical entity in the head and neck region. It is a rare cyst in the oral cavity containing gastric or intestinal mucosa. We report a case of heterotopic gastrointestinal cyst of the tongue in a Nigerian male neonate who presented with a sublingual mass at birth, initial excessive salivation and difficulty with feeding. He was managed conservatively till three months old. Surgical excision was carried out under general anaesthesia and microscopic examination revealed gastric mucosa and parakeratinized stratified squamous epithelial lining along the cyst lining. Recovery was uneventful with no recurrence up to date.

Keywords: Heterotopic Tissue, Congenital; Gastrointestinal; Cyst; Tongue; Infant; Newborn.

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Introduction

Heterotopic gastrointestinal cyst of the oral cavity, was first introduced by Foderl in 1845.¹ Its synonyms include gastric cystic choristoma, entrocystoma and enteric duplication cyst. It can be found all the way through the gastrointestinal tract including the oesophagus, small intestine, pancreas, gall bladder and Meckel's diverticulum. This lesion is very rare and only about 40 cases have been reported in English literature.²

Typical clinical features at presentation include excessive salivation, difficulty with feeding, swallowing, speech or breathing in isolation or combination.^{3,4} The cyst wall typically comprises of routine gastric mucosa of the type seen in body and fundus of the stomach. Histologically, ciliated columnar and stratified epithelium may be admixed with the gastric mucosa and muscularis mucosa. Also, parietal and chief cells may be found as well as pancreatic tissue.^{3,4} Treatment is usually surgical excision. We report a case of heterotopic gastrointestinal cyst of the tongue in a Nigerian neonate.

Case Report

We were invited to review a two day old male singleton neonate at the neonatal ward of the Obafemi Awolowo University teaching hospital. He was said to have been born with a sublingual swelling which prevented him from being breastfed. Mother was a 28 year old undergraduate and father, a 32 year old teacher. Pregnancy and family histories revealed nothing significant.

Patient was the product of uneventful spontaneous vaginal delivery, who weighed 3.4kg kilogram at birth. Chief complaints at presentation were unsightly mouth and difficulty with breast feeding (grasping the nipple). However, there were no challenges with suckling, swallowing and respiration. He was fed through nasogastric tube passed by the paediatric unit till presentation.

Examination revealed an otherwise normal neonate with a discrete midline sublingual swelling measuring about 3 X 4cm, filling the mouth, elevating the tongue against the palate (Figure 1a). The overlying mucosa appeared clinically healthy. Palpation revealed a soft, fluctuant, mobile, non-reducible, non-pulsatile and non-tender mass. Aspiration yielded a sterile brown viscous fluid with chronic inflammatory cells seen on microscopy. An impression of sublingual epidermoid cyst was made and differential diagnosis was dermoid cyst.

Further examination revealed neonate's ability to suck clinician's gloved little finger effectively. He was commenced on feeding with cup and spoon and soon graduated to breastfeeding with specified convenient nipple positioning. Ultrasound scan revealed a thick-walled cystic mass with fluid levels which increased in size from 3.1 x 2.05cm at presentation to 6 x 5cm by third month (preoperatively). At three months, the patient weighed 6.25kg, was carefully assessed and considered fit to undergo cyst enucleation under general anaesthesia with a working diagnosis of epidermoid cyst. He had a

successful naso-tracheal intubation at first attempt, uneventful cyst enucleation, smooth recovery from anaesthesia and surgery. Lesion was approached through a transverse sublingual incision (Figure 1c). Careful blunt dissection was carried out to raise the oral mucous membrane overlying the cyst lining superiorly. The other aspects of the lesion were exposed with an extension of blunt dissection anteroposteriorly and superoinferiorly. The cyst was delivered in one intact piece. Surgery site was thoroughly irrigated with copious normal saline and re-examined before eventual closure in layers.

Highlights of post-operative care include prophylactic antibiotic regimen (intravenous cefuroxime 200mg and metronidazole 100mg 8hourly for 72 hours both converted to syrup at discharge from hospital for 5 days, analgesic regimen (intravenous paracetamol 75mg 6 hourly for 48 hours subsequently converted to syrup 8hourly pro re nata (as needed) for 72 hours), anti-inflammatory agent (dexamethasone 1mg 6 hourly for 48hours then 0.5mg 12hourly for 48 hours) and feeding through nasogastric tube for 4 days post operatively. Thereafter feeding per oral and gentle oral toileting with cotton wool swabs was instituted. Minimal post-operative oedema with drooling of saliva was observed, but these resolved gradually over time. He was discharged home on the 5th postoperative day. Figures 1e, 1f, 3a and 3b show his 3rd week and 24th month postoperative pictures.

Histological section showed a cavity lined in an area by Para-keratinized stratified squamous epithelium while other areas were lined by a typical gastric mucosa exhibiting gastric pits and lined by a simple columnar epithelium. Seen beneath this epithelium are tubular glands consistent with cardiac glands. Also seen within the cystic wall which is composed of collagenized fibrous connective tissue stroma are bundles of striated muscle fibres, vascular channels containing extravasated red blood cells and some areas of haemorrhage Figures 2a, 2b and 2c. A histologic diagnosis of Heterotopic Oral Gastrointestinal cyst was made.

Discussion

Heterotropic gastrointestinal cysts are abnormally placed rests of gastrointestinal mucosa outside the stomach. Although these lesions are found along the entire digestive tract, oral involvement is extremely

rare while it is most commonly found in the esophagus.⁴ In the oral cavity the ventral surface of the tongue extending to the floor of the mouth is the most common location.³ Heterotopic gastrointestinal cysts of the oral cavity have been described in the literature by several nomenclatures such as lingual foregut duplication cyst, enterocystoma and choriomatous cyst.⁴

Other areas in the digestive tract that have been affected include the duodenum^{6,7}, gallbladder⁸, jejunum⁹, ileum¹⁰, rectum¹¹ and anus.¹² It has also been surprisingly reported in the umbilicus.¹³ Outside the digestive tract, it was reported on the lips, larynx, submandibular glands, epiglottis and anterior neck.¹⁴ Although its definitive aetiopathogenesis is unclear, some theories have been proposed. The most commonly held theory is that of the misplaced endoderm from the primordial stomach during the 4th week of intrauterine life when it is located in the mid-neck region.¹⁵ This theory explains why the heterotopic gastrointestinal cyst of the oral cavity occurs in the anterior two-third of the tongue and floor of the mouth. Other theories proposed that the lesions could arise from island of endoderm lining the primitive stomodueum that became entrapped during fusion of the embryonic process.¹⁶ They could also arise from undifferentiated endoderm subjected to inductive influences¹⁷ or from thyroglossal duct and salivary retention cysts.¹⁸

Prenatal diagnosis through ultrasonography is a possibility¹⁹ and this is usually an indication for elective delivery in a tertiary facility as a precaution to ensure safe delivery. Although most cases have been reported in infants and young children¹ and average age at presentation is put at 10 years, it may range from age 0 (as in the present case) to 60 years. To the best of our knowledge, this case is one of the few presenting at birth and possibly a first Nigerian case in literature. A male predilection (M:F:3:2) has also been reported.³

Although mother had antenatal ultrasonography, failure to detect the cyst can be attributed to size of the cyst at last ultrasonography, the skill of radiologist and facilities available for the radiologist. While most patients are reportedly asymptomatic, some may present with difficulty in feeding. Other possible symptoms include difficulty in

swallowing, speech and respiratory compromise.²⁰

These features are also typical of our listed provisional and differential diagnoses. The rarity of heterotopic gastrointestinal cyst of the tongue was the only reason for its exclusion on our list. Our provisional and differential diagnoses actually share the

same clinical features as presented in this case. While a difference between the epidermoid and dermoid cyst can be determined by the presence or absence of skin appendages, (i.e. epidermoid cyst has a lining of epidermis without skin appendages, the dermoid cyst has accompanying skin appendages) histologically.



Figure 1: The clinical frontal view of the patient at birth (a), preoperatively (b), with surgical incision marking (c) and post excision site (d). The Patient at two weeks (e) and twenty fourth month post-operatively (f).

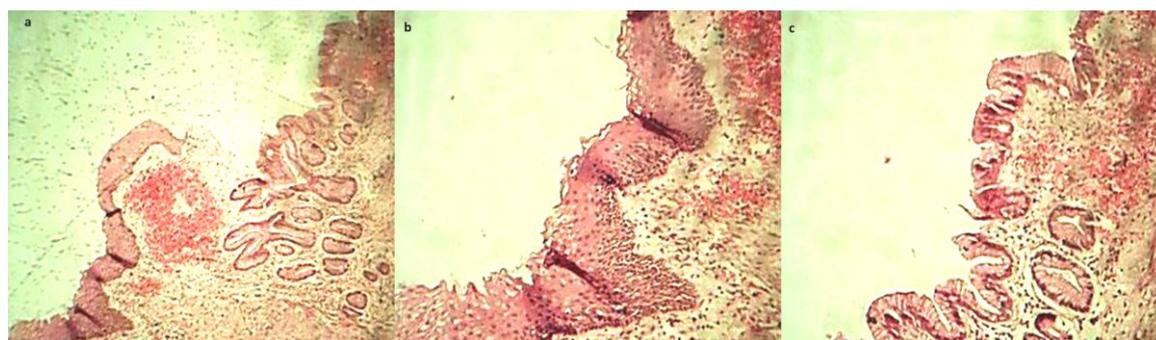


Figure 2: The low power photomicrograph showing a representative section of heterotopic oral gastrointestinal cystic cavity lined by keratinized stratified squamous epithelium (left) and gastric mucosa (right) (H&E x10) (a). The high power view of the same section confirming parakeratinized stratified squamous epithelium (H&E x40) (b) and gastric mucosa exhibiting the gastric pits and cardiac glands (H&E x40) (c).

Due to the rarity of this lesion optimal management is still not known, however complete excision is most favoured. Initial management of symptomatic cases depend largely on the pattern of presentation. Aspiration of the cyst could give temporary

relief as demonstrated in this case where after aspiration of the lesion the patient could suck effectively. Although there may be need for aspiration to decompress the cyst and achieve relief from symptoms, this should be seen as a temporary emergency

measure because of the risk of infection.¹ In this case, aspiration of the lesion was performed twice under aseptic condition and antibiotics to cover. The first was done to investigate the nature of aspirate while the other was done to relieve the child of what the parents described as worsening symptoms.

Our decision for definitive surgery at three months was predicated on the need to have the child attain a weight and status that could be considered more suitable for withstanding the planned procedure under general anaesthesia and facilitating tissue handling. Although it is possible to argue that the cyst could actually serve as a hindrance to feeding and growth, the child in this case gained weight steadily and quite remarkably. If he failed to feed properly per oral and thrive, we could have resumed nasogastric tube feeding and carried out his definitive surgery much earlier. An impending respiratory embarrassment could be another reason for prompt intervention with necessary precautions regardless of patient's weight or age. The cyst lining at surgery was quite firm and distinct from surrounding tissue thus facilitating its easy dissection. Histological section showed a cavity lined in an area by parakeratinized stratified squamous epithelium while other areas are lined by a typical gastric mucosa exhibiting gastric pits and lined by a simple columnar epithelium. Seen beneath this epithelium are tubular glands (consistent with cardiac glands). These glands were also seen within the cystic wall which is composed of collagenized fibrous connective tissue stroma and bundles of striated muscle fibres, vascular channels containing extravasated red blood cells and some areas of haemorrhage Figures 2a, 2b and 2c. A histologic diagnosis of Heterotopic Oral Gastrointestinal cyst was made. Histologically, heterotopic gastrointestinal cysts of the oral cavity are lined by gastric or intestinal epithelium that can be found together with stratified squamous epithelium (as seen in this case) or ciliated columnar epithelium or even simple columnar epithelium.²¹ In most cases smooth muscle is present in the cyst wall.^{1,4}

Aside from the routine prophylaxis for postoperative infection, we employed anti-inflammatory agent because of anticipated postoperative oedema and its unwanted consequences in tongue / floor of the mouth. Incomplete excision could result into

recurrence after many years but very uncommon if total excision was done.¹⁸ Also recurrence has been associated with intraosseous component of the cyst¹⁴ therefore it is advisable to ascertain extent of the cyst by Computed Tomography and or Magnetic Resonance Imaging before surgery but our patient could not afford either of these. Total excision of the cyst was however performed in this case. Most published case reports suggest that majority of patients present relatively later than our patient did. Treatment like in our case was mainly complete surgical excision and no occasion of recurrence has attended reported cases.

Conclusion

Heterotopic gastrointestinal cysts of the oral cavity are rare lesions. Most of these lesions present in infants and early childhood, some may become symptomatic in adulthood. While optimal management is still debated, complete surgical excision is recommended as demonstrated in this case.

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References

1. Barnes L. Surgical pathology of the head and neck. 2nd Ed. New York: Marcel Dekker; 2001:1481-3.
2. Zabolinejad N, Hiradfar M, Khadivi E, Gharvi M. Heterotopic Oral gastrointestinal cyst : Report of two cases Iranian Journal of Otorhinolaryngology 2010;22:151-4.
3. Coric M, Seiwert S, Bumber Z. Congenital oral gastrointestinal cyst: An immunohistochemical analysis. Eur Arch Otorhinilaryngol 2000;257:459-61.
4. Eaton D, Billings K, Timmons C, Booth T, Biavati JM. Congenital foregut duplication cyst of the anterior tongue. Arch Oto Head Neck Surg 2001;127:1484-7.
5. Melato M, Ferlito A. Heterotopic gastric mucosa of the tongue and oesophagus.

- ORL J Otorhinolaryngol Related Spec 1975;37:244-54.
6. Kabria R, Butt S, Ali SA, Akram S. Heterotopic gastric tissue in duodenal bulb mimicking duodenal cancer: an unusual duodenal lesion. *J Gastrointest Cancer* 2009;40:142-3.
 7. Mann S, Mann SK, Rachute E. Heterotopic gastric tissue in the duodenal bulb. *J Clin Gastroenterol* 2000;30:303-6.
 8. Hayama S, Suzuki S, Takahashi S, Hazama K, Fujita M, Kondo S, Katoh H. Heterotopic gastric mucosa in the gallbladder; report of two cases. *Surg Today* 2010;40:783-7.
 9. Boybeyi O, Karnak I, Gucer S, Orhan D, Senocak ME. Common characteristics of jejunal heterotopic gastric tissue in children: a case report with review of the literature *J Pediatr Surg* 2008;43:19-22.
 10. Chan GS, Yuen ST, Chu KM, Ho JW, Leung SY, Ho JC. Helicobacter pylori in Meckel's diverticulum with heterotopic gastric mucosa in a population with relatively high H. pylori prevalence rate. *J Gastroenterol Hepatol* 1999;14:313-6.
 11. Garmendia M, Aparicio JR, Martinez J, Casellas JA. Heterotopic gastric mucosa in the rectum. *Gastrointest Endoscopy* 2007;65:921-25.
 12. Steele SR, Mullenix PS, Martin MJ, Ormseth E, Weppeler E, Graham J, Place RJ. Heterotopic gastric mucosa of the anus: a case report and review of the literature. *Am Surg* 2004;70:715-9.
 13. Heo YS, Jeong SY, Son SW, Kim IH, Seo SH. Heterotopic gastric mucosa in the umbilicus. *Ann Dermatol* 2010;22:223-5.
 14. Noorchashm N, Huff DS, Bartlett S. A mixed heterotopic gastrointestinal cyst of the oral cavity with an intraosseous component. *Plast Reconstr Surg* 2004;114:1195-7.
 15. Toker R, Maddalozzo J, Chou P. Sublingual enteric duplication cyst. *Arch Path Lab Med* 2000;124:614-5.
 16. Khunamornpong S, Yousukh A, Tananuvat R. Heterotopic gastrointestinal and pancreatic tissue of the tongue. A case report. *Oral Surg Oral Med Oral Path Oral Radiol Endod* 1996;81:576-9.
 17. Woolgar JA, Smith AJ. Heterotopic gastrointestinal cyst of oral cavity: A developmental lesion? *Oral Surg Oral Med Oral Path Oral Radiol Endod* 1988;66:223-5.
 18. Martone CH, Wolf SM, Wesley RK. Heterotopic gastrointestinal cyst of the oral cavity. *J Oral Maxillofac Surg* 1992;50:1340-2.
 19. Paladini TM, Guid F, Lambetti A, Martinelli P. Ultrasound. *Obstet Gynecol* 1998;11:141-3.
 20. Morgan WE, Jones JK, Faitz CM, Hicks MJ. Congenital Heterotopic gastrointestinal cyst of the oral cavity in a neonate: Case report and review of literature. *Int J Pediatr Otorhinolaryngol* 1996;36:69-77.
 21. Said-Al-Naif N, Fantasia JE, Sciubba JJ, Ruggiero S, Sachs S. Heterotopic oral gastrointestinal cyst: report of two cases and review of literature. *Oral Surg Oral Med Oral Path Oral Radiol Endod* 1999;88:80-6.

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