J Anesth (2012) 26:438–441
DOI 10.1007/s00540-012-1334-5

CLINICAL REPORT

Anesthetic management of a neonatal lingual gastric duplication cyst: report of a rare case

Harprit Kaur Madan · Lipika Swain · Jyoti Borkar

Received: 22 August 2010 / Accepted: 15 January 2012 / Published online: 21 February 2012

Abstract Gastrointestinal duplications can occur anywhere from the mouth to the anal cavity. The occurrence of a duplication cyst in the tongue of a neonate is very rare. These cysts occur due to endodermal cells that are trapped during development. Congenital lingual cystic masses are challenging entities. They usually present in the neonatal period and surgical excision is curative. We report a 10-day-old, 3-kg neonate with a gastric duplication cyst in the oral cavity with inability to close his mouth or breast feed for whom we performed subtotal excision of the cyst.

Keywords Lingual gastric duplication · Enterocystoma · Foregut duplication cyst

Introduction

Alimentary tract duplications are rare congenital anomalies that can present diagnostic and therapeutic difficulties. These entities were first reported by Duncan and Daniel in 1942. Foregut duplications occurring in the tongue are quite rare, with very few cases reported in the literature. Lingual foregut duplications arise from endodermal cells that become trapped during the process of the fusion of the lateral lingual swelling (distal tongue bud) and the tuberculum impar (median tongue bud), in a 3- to 4-mm embryo. Various synonyms do exist, such as enterocystomas, choristomas, and gastric heterotropia [1]. These masses can seriously compromise the airway, potentially causing hypoxia and death if not recognized and managed appropriately. Prenatal sonographic diagnosis of these congenital anomalies permits (a) anticipation of an airway problem at the time of delivery and (b) the formulation of an algorithm for airway management while oxygen delivery to the baby is maintained through the placental circulation [2]. We present a 10-day-old 3-kg neonate with a gastric duplication cyst in the oral cavity with inability to close his mouth or breast feed for whom we performed subtotal excision of the cyst.

H. K. Madan (✉) · L. Swain · J. Borkar
TN Medical College & BYL Nair Ch. Hospital, Mumbai, India
e-mail: drpreet@hotmail.com

Case report

A 10-day-old neonate, weighing 3 kg, presented with swelling in the oral cavity associated with difficulty in feeding and closing his mouth. On examination, the child was alert, crying well with moderate general condition, and with no other congenital anomaly. Cardiovascular and respiratory systems were normal. The baby's mouth was wide open with the tongue protruding outside due to a 7 × 6-cm, cystic, non-transilluminant mass occupying the entire oral cavity (Figs. 1, 2) The origin of the swelling was difficult to ascertain, but it was occupying the ventral aspect of the tongue and pushing the dorsum of the tongue upwards towards the palate. Clinically, a provisional diagnosis of mucus retention cyst or lymphangioma was made. Preoperative sonography of the swelling revealed a multiloculated cystic swelling with septations, probably suggestive of lymphangioma or a ranula. X-ray of the skull showed a soft-tissue swelling in the oral cavity (Fig. 3). Surgical excision was planned, as the surgeons were not keen on aspirating the cyst. High-risk informed consent and tracheostomy consent were obtained. Ear, nose, and throat (ENT) surgeons were kept on standby during induction. Preoxygenation was tried with a bigger size (size − 1) mask instead of size 0 but was not very effective. Thus, awake

oral intubation was planned. After adequate preoxygenation was achieved, the oral swelling was depressed gently with a malleable retractor and the laryngoscope blade was accommodated. Laryngoscopy was performed with a Miller blade 00. Optimal external laryngeal manipulation (OELM) was applied. An uncuffed red rubber endotracheal tube no. 3 was introduced by an assistant, and after confirming of the tube position by auscultation, capnometry, and chest movements, a Jackson Rees pediatric circuit was attached and balanced general anesthesia was given using O_2, N_2O, sevoflurane (1.5%) and i.v. Atracurium 1.5 mg. A nasogastric tube was inserted. The throat was packed. I.V Dexamethasone 5 mg and i.v. Paracetamol 15 mg were administered. Blood loss was 30 ml and was replaced with 30 ml of blood transfusion. Submucosal excision of the cyst was done. Intraoperative parameters were within normal limits. Tongue stitch was taken. Patient was reversed with i.v. Neostigmine 0.15 mg and Atropine 0.1 mg and extubated after adequate return of reflexes and normal tidal respiration. A number 2.5 polyvinyl chloride endotracheal tube was used as a nasopharyngeal airway. Postoperatively the child was kept in an oxygen tent. The nasopharyngeal airway (number 2.5 polyvinyl chloride endotracheal tube) and the tongue stitch were removed on the 3rd postoperative day. The child was shifted to the ward on postoperative day 4, with no complications. Histopathology showed the cyst wall with columnar epithelial lining of the gastric fundic variety, with smooth muscle and

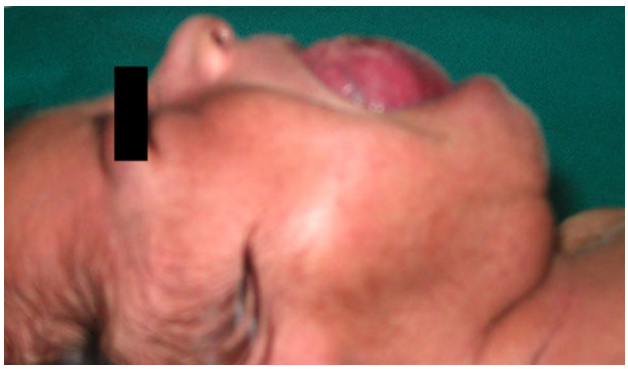

Fig. 1 Lateral profile of neonate with the intraoral cyst

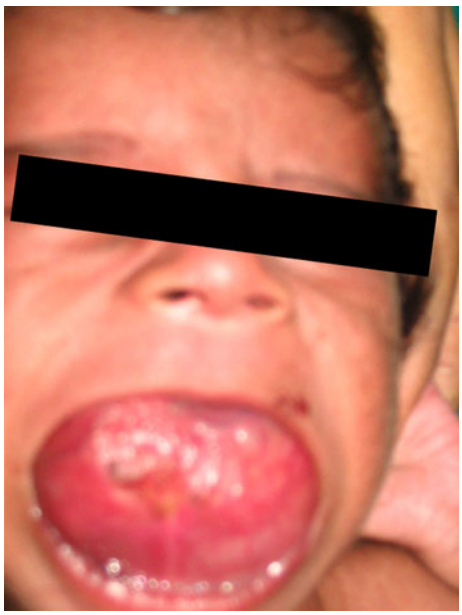

Fig. 2 Photograph showing the neonate with open mouth due to the lingual cyst

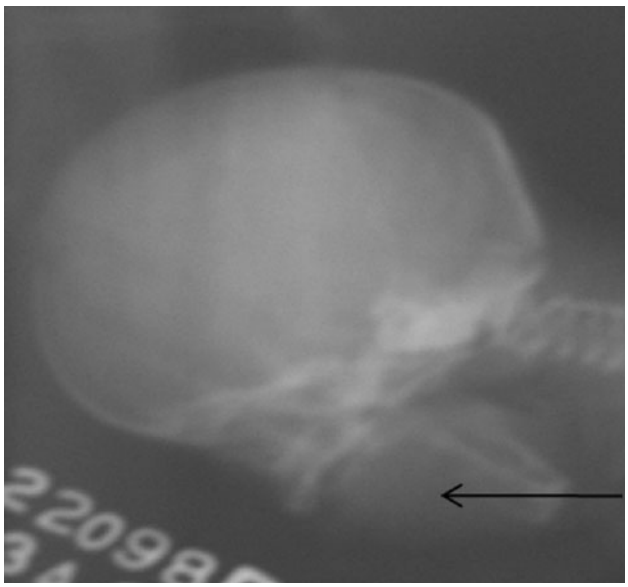

Fig. 3 X-ray of the skull, showing soft-tissue swelling in the oral cavity

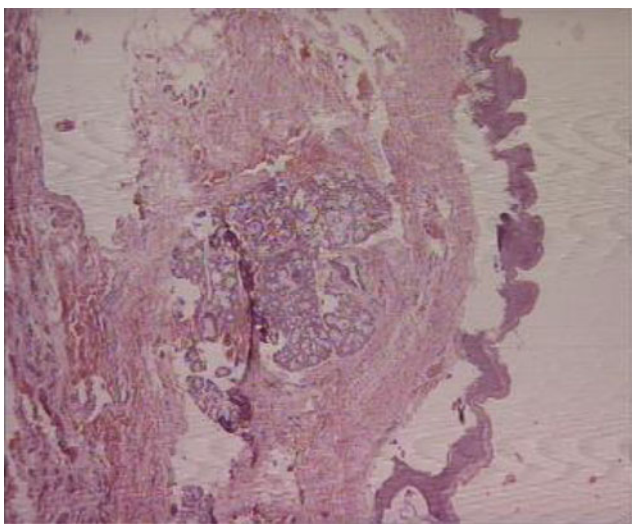

Fig. 4 Histopathology under high power (40×) showing columnar gastric epithelium of the fundic variety and smooth muscle

connective tissue, suggestive of a gastric duplication cyst (Fig. 4). A diagnosis of enteric duplication cyst was established.

Discussion

Gastrointestinal duplication is a true congenital anomaly. Alimentary tract duplications are cystic or tubular structures, lined by normal gastrointestinal mucosa. The criteria for duplications are—they are firmly attached to part of the alimentary tract, have a well-developed blood supply, often shared with the normal viscus, and their epithelial lining always resembles some part of the alimentary tract, most often, the adjacent viscus [1]. They are seen in the stomach, thoraco-abdomen (which is the most complicated, with a high mortality rate), in the ileum (which is commonest), in the duodenum, and in the tongue (which is rare) [3].

The differential diagnosis of an anterior tongue swelling are mucus retention cyst, ranula, cystic hygroma, rhabdomyosarcoma, teratoma, hamartoma, hemangioma, and Beckwith–Wiedemann syndrome, a rare multigenetic disorder associated with dysregulation of the expression of genes involving growth and the cell cycle, can result in macroglossia associated with exomphalos, gigantism, and hypoglycemia [4]. Duplication cysts of the gastrointestinal tract are very rare (0.2%) and can be potentially threatening if present in the oropharynx. Airway obstruction is more likely if an ultrasound diagnosis of a head or neck mass is associated with polyhydramnios, probably because of the inability of the fetus to swallow as a result of esophageal and tracheal compression. Extension of the fetal head in utero may also indicate airway compromise, the mass limiting head flexion. In congenital high airway obstruction syndrome (CHAOS) [5], caused mainly by laryngeal stenosis, prenatal ultrasound demonstrates a dilated trachea, large echogenic lungs caused by filling with water, flattened or inverted diaphragm, and ascites [5].

If a duplication cyst is present in the oral cavity, then the clinical strategy should be to assess its consistency, extent, mobility, and space for instrumentation. After inhalational induction, if ventilation is possible then anesthesia can be deepened to perform laryngoscopy, and once the vocal cords are visualized, a muscle relaxant can be given or else a laryngeal mask airway (LMA) can be inserted laterally, after aspirating the swelling. If these strategies fail then surgical airway access can be thought of or the child can be awakened [6].

At the Royal Hospital for Sick Children, Yorkhill, Glasgow, Scotland, only 21 children were diagnosed with a duplication cyst of tongue in the last 42 years, and 9 of the children had other congenital anomalies, which were mostly vertebral anomalies. Only one case was diagnosed antenatally. Diagnosis was arrived following chest X-ray, barium swallow, ultrasonography (USG), and at surgery. Thus, clinicians should have a high index of suspicion in children with an intra-oral cyst and with unusual respiratory and gastrointestinal symptoms [7].

At the Children's Medical Centre, Dallas, Texas (1990–2000), in a retrospective study, 6 children were diagnosed with foregut duplication cysts of the tongue for which they underwent excision. No patients had presented with respiratory compromise despite the large size of the anterior tongue masses (1.5–2.4 cm). Many of these cysts were misdiagnosed as dermoids. Preoperative evaluation includes palpation, high-resolution USG, computed tomography (CT), and magnetic resonance imaging (MRI). High-resolution USG is the investigation of choice as it does not require sedation [2]. If the diagnosis is known antenatally signs of congenital high airway obstruction (CHAO) should be looked for. If CHAO is present then a multidisciplinary approach by the pediatric anesthesiologist, obstetrician, ENT specialist, pediatric surgeon, and neonatologist is needed [4].

Neck masses in the fetus may cause asphyxia during delivery. The survival rate without intervention at birth is 0–20%. If a neck mass is detected in utero, then a strategic plan should be developed [6]. Ex-utero intrapartum treatment (EXIT procedure) or the OOPS procedure (operation on placental support) can be planned. With the EXIT or OOPS procedures, cesarean section is performed under general anesthesia using a halogenated inhalational agent to relax the uterus. The fetus is either fully or partially delivered and the feto-placental circulation is maintained without division of the umbilical cord until the airway is secured. The uterus is relaxed with tocolytics to prevent premature placental separation and to maintain feto-placental flow and oxygen delivery [8].

In our case the baby cried immediately after birth and the majority of the mass was in the anterior part of the tongue, pulling it forward and assisting the airway patency. The decision to perform awake laryngoscopy was made as inhalational induction, though a safer option, might have proven technically difficult for a potentially obstructed airway because of inability to mask ventilate over the protruding tongue. Besides, we had tried ventilating with a bigger face mask (size 1), but it was not successful. Intravenous induction was not acceptable as there was a possibility of difficulty in securing the airway and oxygenation if the intubation proved impossible.

Conclusion

To conclude, gastrointestinal duplications are a true congenital anomaly. They are rarely seen in the anterior

tongue. They can compromise the airway causing hypoxia and death if not recognized and managed appropriately. They are easily misdiagnosed in the preoperative period. If diagnosed antenatally, the aim is to emphasize the importance of a multidisciplinary team approach to the delivery of a neonate with suspected airway obstruction, and to carefully consider the options available to maintain oxygenation while securing the airway at delivery. Aspiration of the cyst can be done for adequate access to the airway. Airway achievement is the mainstay of management and preparation for tracheostomy is mandatory.

References

1. El-Bitar MA, Milmoe G, Kumar S. Intralingual foregut duplication cyst in a newborn. Ear Nose Throat J. 2003;82:454–6.
2. Eaton D, Billings K, Timmons C, Booth T, Biavati JM. Congenital foregut duplication cysts of the anterior tongue. Arch Otolaryngol Head Neck Surg. 2001;127:1484–7.
3. Willner A, Feghali J, Bassila M. An enteric duplication cyst occurring in the anterior two-thirds of the tongue. Int J Pediatr Otorhinolaryngol. 1991;21:169–77.
4. Skelton VA, Goodwin A. Perinatal management of a neonate with airway obstruction caused by rhabdomyosarcoma of the tongue. Br J Anaesth. 1999;83:951–5.
5. Hedrick MH, Ferro MM, Filly RA, Flake AW, Harrison MR, Adzick NS. Congenital high airway obstruction syndrome (CHAOS): a potential for perinatal intervention. J Pediatr Surg. 1994;29:271–4.
6. Stocks RM, Egerman RS, Woodson GE, Bower CM, Thompson JW, Wiet GJ. Airway management of neonates with antenatally detected head and neck anomalies. Arch Otolaryngol Head Neck Surg. 1997;123:641–5.
7. Carachi R, Azmy A. Foregut duplications. Pediatr Surg Int. 2002;18:371–4.
8. Skarsgard ED, Chitkara U, Krane EJ, Riley ET, Halamek LP, Dedo HH. The OOPS procedure (operation on placental support): in utero airway management of the fetus with prenatally diagnosed tracheal obstruction. J Pediatr Surg. 1996;31:826–8.