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Case report

Serious neonatal airway obstruction with massive congenital sublingual ranula and contralateral occurrence

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HIGHLIGHTS

- A well considered plan is required when a neonatal surgical airway is required.
- Intra-oral ranulas can be initially managed with marsupialisation or cyst excision.
- Sublingual gland excision is more reliable but associated with higher risk.
- Sublingual gland excision should be reserved for recalcitrant cysts.

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ABSTRACT

Background: Congenital ranulas seldom occur, with bilateral presentation and prenatal diagnosis reported very rarely. We believe this is the first reported case of a neonate with a antenally diagnosed massive congenital ranula, who went on to develop a non-contiguous contralateral ranula, both contributing to obstruction in a complex paediatric airway.

Case report: A female neonate was born to a non-primagravid mother via a planned elective caesarean section due to a lower facial defect and oral cyst. Antenatal aspiration of the pseudocyst was performed under ultrasound guidance with limited success. In the immediate post-natal period a poor airway was observed and the cyst was subsequently marsupialised. With the development of macroglossia secondary to oedema and tongue base collapse the airway was secured through surgical tracheostomy. A subsequent ultrasound scan revealed the presence of a second solitary cystic mass on the contralateral side. After careful excision of the contralateral pseudocyst, tongue function improved, with the resolution of a safe airway which permitted successful decannulation. A planned definitive procedure antenatally did not result in the anticipated improvement in function. However the subsequent development of a second non-contiguous pseudocyst and further surgical management resulted in a safe airway, improved masticator function and the ability to thrive.

Conclusions: The prenatal diagnosis of congenital ranulas have been seldom reported, with no reported cases of contralateral occurrence and airway obstruction from an intraoral ranula. This rare case highlights the need for a well considered contingency plan when surgery is required for a neonatal airway at risk.

1. Background

Congenital ranulas are rare epithelial lined retention cysts or pseudocysts, which arise in the floor of the mouth. The term ranula itself is derived from the Latin word *rana*, meaning frog and ranula describing a little frog; denoting its resemblance to a bulging frog's underbelly [1]. They are thought to occur following obstruction of the main sublingual duct or acini causing extravasation of mucus into the surrounding tissues [2]. Rarely the aetiology is secondary to sublingual gland ductal atresia or failure of embryological canalisation [1].

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Ranulas are classified anatomically. Simple or intraoral ranulas are confined to the sublingual space whilst plunging or cervical ranulas extend via herniation of the pseudocyst through the mylohyoid into the submandibular neck space [2]. Epidemiological data is limited, but the overall prevalence of ranulas in the paediatric population is 0.2 cases per 1000 [3]. Studies have shown that oral ranulas tend to occur in children and young adults and peak in their frequency in the second decade of life [3]. Plunging ranulas tend to occur in the older age group and are more infrequent. Congenital ranulas are rare, with an incidence of 0.74%, with prenatal diagnosis rarely reported [4]. Whilst ranulas can infrequently recur [5], contralateral occurrence of congenital ranulas has not yet been described in the literature.

Most ranulas are asymptomatic and self-limiting, presenting as a painless, bluish cystic swelling in the floor of the mouth. In the submandibular gland, mucus extravasation initiates an inflammatory reaction leading to fibrosis that often prevents further expansion of the pseudocyst. In sublingual gland however, the secretory activity of the gland is persistent and continues despite fibrosis leading to more substantial pseudocyst formation [2]. An extremely small proportion can become large enough to cause airway compromise, feeding difficulties and facial defects [4]. There is no consensus opinion on the management of these lesions and there is often great variation in practice. Multiple options exist, including surveillance, needle aspiration, surgical excision of the cyst, sublingual gland excision along with the cyst, marsupialisation, sclerotherapy, laser excision or cryosurgery [6]. However, whichever treatment is implemented, the aim is to restore normal function and decrease the risk of recurrence with as little morbidity as possible.

We describe a complex case of massive congenital sublingual ranula contributing to airway obstruction with post intervention occurrence of a second non-contiguous contralateral ranula.

2. Case presentation

A female neonate was born to a non-consanguineous primagravid mother at 37 weeks gestation. Delivery was by planned elective caesarean section for an antenatally diagnosed lower facial defect and oral cyst. (See Fig. 1) There were no other complications during the pregnancy and no significant family history.

Antenatal aspiration of the pseudocyst under ultrasound (US) guidance was attempted. Although 20 ml of white gelatinous fluid was aspirated, the outcome was not one of significant clinical improvement. No other cystic structures were noted at this time on US examination. The pseudocyst persisted after birth resulting in a poor oral airway in the immediate post-natal period, the presence of stertor prompted endoscopic evaluation of the upper aerodigestive tract under a general anaesthetic, which demonstrated mild epiglottitis. The extent of the oral pseudocyst was determined by transillumination and subsequent marsupialisation permitted safe oral intubation.

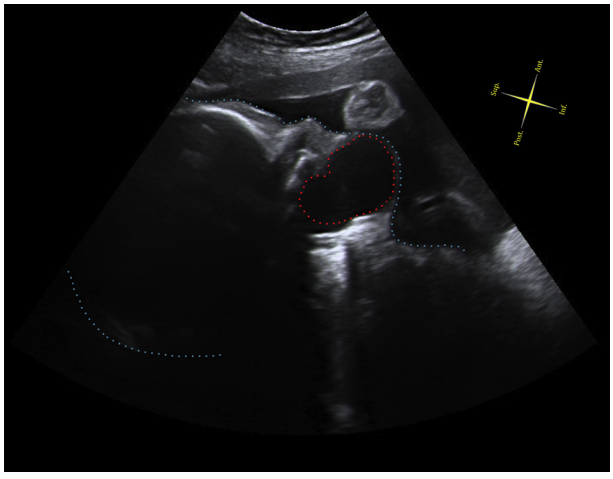

Fig. 1. Antenatal U/S at 37/40 of fetal head. Demonstrating a large intraoral cyst highlighted by red broken line. Blue broken line outlines fetal head.

Post operatively, the neonate developed a right-sided tension pneumothorax, which was managed with two intercostal chest drains; the child remained intubated. By the third post-operative day, the floor of the deroofed pseudocyst was epithelializing with no sign of infection, although a floor of mouth mass persisted. Clinical examination revealed glossal and masticator hypotonia in the presence of micrognathia. Ultrasound imaging of the tongue and floor of mouth was performed excluding any residual cystic element. Corticosteroid treatment was subsequently commenced.

After the first post-operative week the tongue swelling had significantly reduced prompting a trial extubation in theatre. Direct laryngo-tracheoscopy revealed no abnormality however on extubation, significant tongue base collapse was observed causing considerable upper airway obstruction and persistent desaturation. Conservative measures with the use of airway adjuncts to improve the airway did not help. A decision was made to proceed to a surgical tracheostomy to secure the airway. Post-operatively there were no further airway concerns, however oral feeding remained a challenge and nasogastric feeds were implemented.

At seven weeks old, further US imaging of the tongue and floor of mouth was performed which unexpectedly revealed a second non-contiguous sublingual cystic mass ($2.0 \times 3.0 \times 2.5$ cm) on the contralateral side. The mass contained echogenic debris and did not demonstrate peripheral or central vascularity. The differential diagnosis included lymphatic malformations, haemangioma, teratoma, sublingual dermoid cyst and thyroglossal cyst. However a diagnosis of a contralateral ranula was made based on clinical and radiological findings. The pseudocyst was completely excised under general anaesthetic. A further attempt at decannulation was made but was unsuccessful due to ongoing tongue base collapse.

Post-operatively there was marked glossomegaly secondary to oedema, which resolved following a prolonged period of enteral corticosteroids. The neonate progressively became tolerant of an oral diet with supplementary nasogastric nutrition which helped weight gain. Improved mastication and glossal function was observed. At two months of age decannulation was successful and a safe airway achieved. The child continued to thrive, and follow-up at one year showed no evidence of recurrence.

3. Discussion

Symptomatic congenital ranulas are extremely rare, with only one described in the literature [7]. We believe we have described the first reported case of a antenatally diagnosed congenital massive intraoral ranula in a complex airway, that after failed antenatal aspiration and successful postnatal marsupialisation, a second non-contiguous pseudocyst developed on the contralateral side. Both ranulas contributed to airway compromise on a background of micrognathia, glossomegaly and tongue base collapse.

Whilst ultrasound is a mainly dynamic process, retrospective review of the antenatal images did not identify another intraoral cyst. The de novo development of a contralateral ranula is unlikely and we propose that either the large ranula obscured a smaller contralateral developing congenital cyst or that it was simply too small to observe on initial imaging.

Whilst a congenital predisposition to ranula formation has been suggested given the preponderance in the Asian population;

especially in the case of the plunging ranulas; no specific genetic factors have yet to be identified [8,9]. There are numerous reports of congenitally imperforate submandibular glands from failure of duct canalisation causing sublingual cystic swelling via connection through the ducts of Rivinus [10,11]. The submandibular glands develop laterally to the developing tongue as endodermal buds in the median paralingual groove and Wharton's duct forms from closure of the anterior three quarters of this groove. This solid cord then hollows out to form a patent duct, failure of which results in an imperforate submandibular duct [11].

Other hypotheses may be more plausible accounting for the increased incidence in sublingual glands. A prospective study evaluated ranula pathogenesis according to anatomical variation of the sublingual gland [12]. Usually numerous ductules from the posterior sublingual gland open onto the summit of the sublingual fold; however several of the ductules can also join to form a common duct (Bartholin's duct) that empties into Wharton's duct. Through meticulous dissection, they found that 88.9% of simple ranulas contained a Bartholin's duct in comparison to 42.9% of plunging ranulas. This was compared to 0% in control patients.

Ranulas are often misdiagnosed and although their diagnosis is usually a clinical one, the treating clinician should supplement their examination findings with radiological investigations. US, computed tomography and fine needle aspiration have been shown to be useful when narrowing the potential differentials [6]. Management of ranulas is a polarising topic, with conflicting evidence as to which treatment modality is best. Optimal management is initial observation to avoid trauma. However in our patient, the presence of airway compromise and feeding difficulty necessitated definitive surgical management. Whilst sclerotherapy, cryotherapy and laser excision have both demonstrated success in some studies, the majority of treatment is based on either simple marsupialisation or excision of the ranula with or without the sublingual gland [2].

In a review of 580 ranulas, recurrence rates for marsupialisation, excision of ranula and excision of the sublingual gland or gland combined with the lesion were 66.7%, 57.7% and 1.2%, respectively [3]. Whilst effective at minimising recurrence risk, morbidity of sublingual sialadenectomy is relatively high with risk of injury to Wharton's duct (2%), bleeding (1–2%), infection (1–2%) or lingual nerve paraesthesia (2–12%) [13].

Baurmash advocates a conservative approach through marsupialisation and positive pressure packing of the deroofed cavity. This initially temporarily seals the ductal leak and later stimulates a significant inflammatory reaction for fibrosis to permanently seal the leak [14]. This method is informed by the relatively high rate of misdiagnoses of floor of mouth swellings that are often unrelated to the sublingual gland. These include cystic lesions at Wharton's duct orifice and mucoceles arising from the incisal gland in the anterior floor of mouth. This method of packing may reduce recurrence rates to 10% however the results remain unpredictable and several days of packing can be distressing [13,14]. McGurk therefore proposes marsupialisation with packing as the initial treatment option with sublingual gland excision for persistent recalcitrant lesions [13]. As in our case, excision of the cyst without the sublingual gland, whilst carrying a higher recurrence rate reduces the risk of complication, which may have greater morbidity in infants.

4. Conclusion

A planned definitive procedure antenatally did not result in an improvement in function that one would expect. However the subsequent development of a second non-contiguous pseudocyst and further surgical management resulted in a safe airway, improved masticator function and the ability to thrive. The prenatal diagnosis of congenital ranula, which becomes symptomatic has seldom been reported; with no other cases describing a subsequent contralateral occurrence after initial treatment. This rare case highlights the need for a staged and thorough investigation of floor of mouth swellings with consideration of definitive sublingual gland excision if recurrence after initial successful marsupialisation or cyst excision. Importantly, when managing the neonatal airway the clinician should always have a contingency plan in place, involving the multi-disciplinary team, including the otolaryngologist, anaesthetist, paediatrician in this complex group of patients.

Conflict of interest

None.

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Ethical approval

Written consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in Chief of Annals of Medicine and Surgery.

Author contribution

MMG led the case write up and literature review overall. OM participated in the management of this patient, gained consent for use of images for publication of this case, advised on write up and edited the manuscript. KS lead initial drafts of the entire manuscript. JG made large contributions to drafting this manuscript several times. MR led the care of this patient, advised on themes to cover for drafting and edited the manuscript. All authors have read and approved the final manuscript.

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Not applicable.

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