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Case report of prenatal diagnosis and surgical treatment of congenital ranula

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ABSTRACT

Congenital cystic pathologies of the mouth are very rare. The term "ranula" describes a swelling in the floor of the mouth, caused by a mucous extravasation cyst (MEC) or, less commonly, a mucous retention cyst (MRC); it derives from the main sublingual or submandibular salivary glands. We describe a case of congenital mucous cyst of the mouth's floor, diagnosed in-utero by ultrasound in a fetus at 27 weeks' gestation. Ultrasound scan follow-up revealed no changes in the size or position of the cyst. The fetal growth was normal, as the amniotic fluid volume. Surgical treatment was performed five days after delivery. There were no complications and no recurrences occurred to now. The diagnosis was confirmed by postnatal histology.

SOMMARIO

Le patologie cistiche congenite della bocca sono molto rare. Il termine "ranula" descrive un gonfiore nel pavimento della bocca, causato da una cisti da stravasamento di muco (MEC) o, meno comunemente, da una cisti da ritenzione di muco (MRC); deriva dalle principali ghiandole salivari sub-linguali o sottomandibolari. Descriviamo un caso di cisti mucosa congenita del pavimento della bocca, diagnosticata in utero tramite ecografia in un feto alla 27^a settimana di amenorrea. Il follow-up ecografico non ha rivelato cambiamenti nelle dimensioni o nella posizione della cisti. La crescita fetale era normale, come il volume del liquido amniotico. Il trattamento chirurgico è stato eseguito cinque giorni dopo il parto. Non ci sono state complicazioni né recidive avvenute fino ad ora. La diagnosi è stata confermata dall'esame istologico postnatale.

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INTRODUCTION

Congenital cystic pathologies of the mouth are very rare. The term "ranula" describes a swelling in the floor of the mouth, caused by a mucous extravasation cyst (MEC) or, less commonly, a mucous retention cyst (MRC); it derives from the main sublingual or submandibular salivary glands (1). MECs develop by disruption of minor salivary gland ducts and subsequent extravasation of mucous secretions into the contiguous connective tissue, whereas MRCs develop by proximal expansion of a blocked duct. MECs mostly occur in children and young adults, rarely in newborns. The main difference between the two abnormalities is that, differently to MECs, MRCs are lined by epithelium. The incidence of congenital ranula is about 0.74% (2).

Clinically the fluid-filled pseudocyst elevates the tongue. These pseudocysts are normally sited in the sublingual space between the mylohyoid muscle and the lingual mucosa. Occasionally, the edema produced by its growth extends into the submental or submandibular space through the mylohyoid muscle. The differential diagnosis of ranula are lymphatic malformations and fetal cervical tumors, such as teratomas, thymomas and thyroid tumors (3). An appropriate diagnosis is extremely important, as treatment and prognosis of both conditions are quite different (4). International scientific literature reports only few cases of prenatal diagnosis of ranula. We describe a case of congenital mucous cyst of the mouth's floor, diagnosed in utero by ultrasound in a fetus at 27 weeks' gestation, and confirmed by postnatal histology. This case report describes also the postnatal surgical treatment of this abnormality. The main novelty of this case report is represented by the fact that an accurate ultrasound prenatal diagnosis based on the size of the ranula allows us to know in the antenatal period (and confirm after birth) that was not necessary to perform an ex utero intrapartum treatment (EXIT) procedure, and we could wait five days for surgical operation.

CASE REPORT

A 29-year-old Caucasian woman, gravida 1, para

0, with a history of two previous surgical treatments for truncus arteriosus, came to our observation to perform a routine second-trimester fetal ultrasound scan at 27 weeks' of gestation. The scan showed a single viable intrauterine fetus with a hypoechoic mass in the floor of the mouth. The cyst was sited below the tongue and ahead to the inferior part of the maxillary bone. It measured 28 mm (length) and 21 mm (width). The margins were clear and no echoes could be observed inside the mass (**Figure 1-2**).



Figure 1. Ultrasound diagnosis of ranula



Figure 2. Ultrasound diagnosis of ranula

The tongue was displaced upwards, but all other findings were within normal limits. The tongue moved normally and the fetus appeared to swallow normally. Color Doppler imaging showed no neovascularization, either inside or outside the cyst.

She had run the first trimester screening for aneuploidies with negative results and even after the diagnosis of sublingual cyst, she decided to not perform amniocentesis.

Two follow-up ultrasound scans were performed at 31 weeks and at 36 weeks of gestation, and no changes were observed. The fetus showed normal development, the cyst did not change in size or consistency and there was no evidence of polyhydramnios. Cesarean section was performed at 40 weeks' gestation for premature rupture of membranes and fetal breech presentation. A normal 2900 g female baby was born with an Apgar score of 9 at 1 min and 10 at 5 min. The infant had a 40 mm sublingual cystic mass (**Figure 3**) which displaced the tongue upwards.

Her gag reflex was normal. Infra buccal and cervical prolongations were not found. Five days after delivery, a cyst's marsupialization was performed, and a new excretory duct of the gland was surgically created.



Figure 3. Surgical treatment of Ranula

The cyst was surgically drained under general anesthesia (**Figure 4-5**). The fluid obtained was mucus-like and the final pathology was identified as a mucosal retention cyst. After 36 hours, the baby quickly resumed a normal breastfeeding and his aesthetic appearance was satisfactory. The patient was discharged 6 days after the surgical operation. At the discharge, a soft tissue edema (false Ranula) was still present, but it was solved spontaneously within 7-10 days. After one month, the clinical follow up showed no evidence of recurrence.

The study protocol was approved by the Ethics Committee of the ARNAS Garibaldi Hospital and conformed to the ethical guidelines of the Helsinki Declaration. The woman signed an informed consent before entering the study, and her anonymity was preserved.



Figure 4. Surgical treatment of Ranula



Figure 5. Surgical treatment of Ranula

DISCUSSION

A classification of the ranulas could be done according to their localization. Simple ranulas are sited in the mouth's floor, cervical ranulas in the paracervical area, and plunging ranulas near the superior airway. Plunging ranulas could extend into the floor of the mouth, and the histopathology is the only way to differentiate them by other types. Ultrasonographically they are indistinguishable.

The best surgical treatment is a two steps operation based on marsupialization, and subsequent posterior resection (1). The differential diagnosis of oral cystic abnormalities involves other exophytic congenital lesions. This group includes epignathus, gingival cyst of the newborn, palatal cyst of the newborn, congenital epulis, vascular hamartomas and lymphangiomas. Another pathology that has to be ruled out is the oropharyngeal teratoma, which is an uncommon congenital tumor, associated to significant morbidity and mortality.

These abnormalities distort the orofacial anatomy and often cause respiratory distress at birth. An optimal management of these conditions requires prenatal diagnosis by ultrasound. The teratoma appears as a solid-cystic tumor with mixed areas of hypo- and hyperechogenicity and it is usually found in association with polyhydramnios. Posterior magnetic resonance imaging has to be subsequently performed to confirm the diagnosis.

Finally, the differential diagnosis of cystic pathologies of the tongue includes thyroglossal duct cyst, with an unusual localization in the mouth's floor, congenital abnormalities of the submandibular duct, heterotopic gastric cyst and enterocystoma (3).

All of these conditions are rare, with a similar hypoechogenic ultrasound pattern, in which magnetic resonance imaging could be helpful to differentiate them (4-5).

Large size ranulas can cause airway obstruction with hypoxia at birth and hind the movements of swallowing causing polyhydramnios. In the massive ranulas, when there is the risk of hypoxia caused by an airway obstruction, the so-called ex utero intrapartum treatment (EXIT) procedure is required (6-7).

This procedure, by partial drainage of the cyst, needs to maintain airway before that fetomaternal circulation is interrupted.

The elective treatment of ranulas is the surgical treatment, and it may have different approaches: aspiration of the cyst, gland marsupialization, excision of ranula, and excision of ranula and ipsilateral salivary gland, crioechexesi.

The airway management of the newborns is

extremely difficult to maintain, particularly in case of large ranulas, so it has to be considered when these surgical interventions have to be planned (8). Prenatal diagnosis of a cystic lesion of the oral cavity, if carried out, requires a careful differential diagnosis to determine appropriate management of delivery and perinatal care. The main decision to consider is related to the size of the ranula: in case of bigger ranulas it has to be decided if perform the ex utero intrapartum treatment (EXIT) procedure or wait and make it in a second time (9-10). In our case report the fetal airway were not compromised at birth, as we preannounced by prenatal ultrasound diagnosis, so it was decided to perform the operation after 5 days.

CONCLUSION

Ranula is an unusual finding. Definitive diagnosis is obtained only after histological examination. When a prenatal cyst in the mouth is diagnosed, an adequate follow-up is necessary to assess the size and potential growth of the cyst, as well as all possible associated complications. According to this, the time of surgery has to be decided accurately, as well as the need for immediate action to ensure airway of the newborn after delivery. An accurate prenatal ultrasound diagnosis, based on the size of the cyst (as described in our case report) could be extremely important to orient the time of surgery.

CONFLICT OF INTEREST

The authors report no conflict of interests.

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