

Congenital mucocele of the tongue: a case report and review of the literature

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Mucoceles are benign lesions of the oral cavity that develop as a result of retention or extravasation of mucous material from minor salivary glands. Congenital mucoceles are very rare. These lesions in newborns may interfere with breastfeeding and may even compromise respiratory function. A patient with a congenital mucocele diagnosed by prenatal ultrasound screening showing a cystic lesion of the tongue is presented herein. The physical examination, lesion evolution and imaging are described, together with the surgical management, histopathology and two-year follow-up. Early clinical assessment, differential diagnosis and magnetic resonance imaging allow clinicians to diagnose and treat this rare congenital condition with surgery in early infancy.

Key words: mucocele, congenital, tongue, prenatal diagnosis.

Congenital cystic lesions of the oral cavity such as mucoceles are very rare¹. Prenatal detection of these lesions is important since they may cause breathing problems at birth and interfere with breastfeeding. The final diagnosis is obtained by histopathological examination of the resected specimen after birth. Mucoceles are lesions of the oral mucosa that result from an alteration of minor salivary glands². Their incidence is estimated as 2.4 cases per 1,000 population, frequently occurring in the second decade of life^{2,3}. Mucoceles, especially congenital ones, in the mouth are also called simple cyst or ranula epithelial cyst⁴. The most common location of these lesions is the lower lip⁵⁻⁷. Mucoceles involving the ventral surface of the tongue are very rare, with sporadic cases reported in the world literature^{4,5}, accounting for 2-8% of oral mucoceles³. We describe herein the ultrasound presentation, clinical course and surgical management of an infant, from prenatal evidence of the tongue mucocele up to two years of age.

Case Report

We present the case of a 36-year-old patient

in her third pregnancy, with a history of spontaneous abortion. At week 24, ultrasound imaging revealed a male fetus with a unilocular, anechoic cystic lesion, 35 mm in diameter, with no solid components and well-circumscribed within the tongue (Fig. 1). The pregnancy course was normal. At week 39 of gestation, the mother had a spontaneous, vaginal delivery. The physical examination of the newborn showed tongue protrusion with volume increase of the ventral surface and inability to close the mouth. A needle puncture of the lesion was performed in the delivery room, and mucous fluid (20 ml) was aspirated. As a result of this procedure, the lesion decreased in size, and the newborn had no difficulty breathing and was able to feed orally. Nevertheless, the mass continued to grow gradually (Fig. 2). Magnetic resonance imaging showed a cystic lesion of 3 x 2 cm in diameter on the inferior and anterior portion of the tongue extending to the right of the middle line (Fig. 3). Surgical resection of the lesion was performed under general anesthesia when the newborn was two months old. The surgical procedure consisted of approaching the lesion through the ventral

surface of the tongue, dissecting the lesion by reaching the muscular layer, and then performing a total exeresis of the mass. The pathological analysis revealed that the cyst wall was lined by salivary gland ductal epithelium, showing no atypical cells and confirming the diagnosis of a retention mucocele (Fig. 4). No further complications arose and the immediate course was excellent. There were no swallowing or breathing difficulties, the patient was able to close his mouth normally, and he has remained asymptomatic, with no evidence of lesion recurrence, in the 22-months follow-up.

Discussion

A mucocele is caused by the accumulation of mucoid material in the oral cavity, which results in an apparent, well-circumscribed, fluctuating, painless, and bluish tumefaction². It originates by trauma to the salivary glands, which causes rupture of the excretory duct or of the acinus, releasing saliva into the adjacent connective tissue (extravasation type mucocele), or by obstruction of the duct, due to alteration in saliva viscosity or ductal atresia (retention type mucocele). In this latter case, the cavity is lined by an epithelium^{1,6}. It has low incidence in infants under one year of age (2.7%). Extravasation type mucoceles are the most

frequent, whereas retention type mucoceles are very rare in children, which makes the present case most unusual^{6,8}. Mucoceles of the anterior portion of the tongue arise from the Blandin-Nuhn glands, which are mixed glands (both serous and mucous), and are located on the ventral surface of the tongue^{3,9}. Sporadic cases, mostly of small lesions, have been reported in the literature¹⁰. Congenital mucoceles may be caused by congenital atresia



Fig. 1. Fetal ultrasound screening at 24 weeks of pregnancy. Cystic lesion of the tongue is observed (arrow). A. Sagittal 2D ultrasound image. B. 4D ultrasound image.



Fig. 2. Clinical examination. A. Lesion on the ventral surface of the tongue. B. Newborn baby feeding orally.



Fig. 3. Magnetic resonance imaging. Coronal section: cystic lesion on the inferior and anterior portion of the tongue.

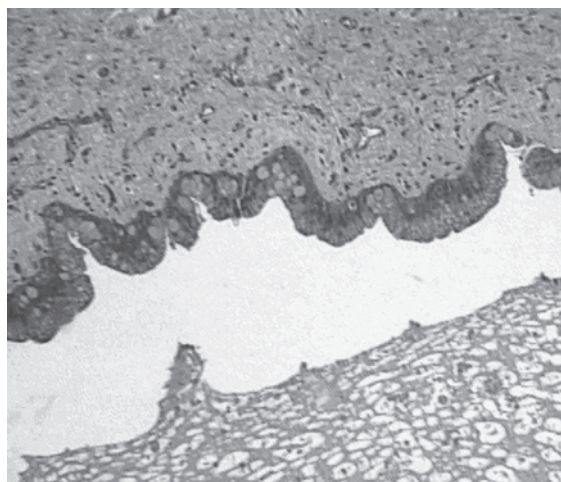


Fig. 4. Histopathology. Cyst has a connective tissue wall, lined by pseudostratified epithelium (hematoxylin-eosin stain, magnification $\times 100$).

of the salivary duct or by trauma to the baby's oral tissues during pregnancy or at birth (intrauterine finger sucking, passage through the delivery channel, use of forceps during delivery, newborn manipulation by attending nurses and doctors)^{2,6}. Congenital masses may cause difficulty in breathing and interfere with breastfeeding⁴. Prenatal diagnosis is fundamental, since it will allow an early detection of respiratory distress to plan an effective management strategy for the prompt establishment of an airway during delivery. Routine follow-up of the lesion size is crucial for managing congenital lesions⁴. The *ex-utero* intrapartum treatment (EXIT) is a specialized surgical delivery procedure used in congenital pathology⁴. This procedure consists in securing fetal oxygenation through maternal-fetal circulation while some procedures are carried out to establish a safe airway in a fetus at high risk of presenting an unsafe or non-patent airway at birth. The baby is partially delivered through a programmed cesarean section. A qualified multidisciplinary team having the adequate training and expertise is required to perform the procedure within a short period of time⁴. In the case of lesions that retain mucus secretion in the oral cavity, a well-planned EXIT procedure involving intrapartum aspiration of the lesion may be effective and life-saving for the newborn baby⁴. To our knowledge, three cases of ranulas (40 mm or larger in diameter) have been successfully treated using this procedure^{1,4,11-13}. In this case, the lesion was located on the anterior portion of the tongue. Both the fetal growth and the amniotic fluid volume were normal, which allowed continuation of the pregnancy until birth without intervention. The ultrasound screening allowed prenatal diagnosis of the cystic lesion, but the final mucocele diagnosis can only be confirmed by histopathological analysis of the specimen following the postnatal removal of the lesion⁴. The differential diagnosis of cystic lesion of the oral cavity includes, among others, epignathus, gingival cyst, palatal cyst, thyroglossal cyst, ranula, congenital epulis, vascular hamartoma, lymphangioma, and oropharyngeal teratoma^{1,4}. In practice, mucoceles are rarely considered a clinical problem. However, their presence in newborns may interfere with breastfeeding and cause parental alarm^{2,6,8}. Mucoceles of the tongue

may easily be traumatized, becoming a source of irritation and discomfort for the patient, therefore requiring prompt removal³. The current standard treatment for these lesions is surgical resection, which is generally advocated when there is difficulty in swallowing, an increase in the lesion size, or doubts regarding their diagnosis². Different surgical techniques have been used for the treatment of mucoceles of the oral cavity, with the technique of choice being the complete surgical excision of the lesion together with the glandular components involved^{6,8,14}. In order to avoid recurrence, the lesion should be removed up to the muscle layer including the small glands found in the surgical field³. Needle aspiration and marsupialization are alternative treatments involving the risk of recurrence¹⁴. Other treatments have been described, including laser ablation, cryosurgery, sclerotherapy, micro-marsupialization, and intralesional corticosteroid injection, among others^{3,5,14,15}. There have also been a few reported cases of spontaneous resolution of these lesions^{4,7,14}. The decision to perform the surgical procedure depends on the extent of the patient's respiration and feeding difficulties as well as on the child's development⁴. An early surgical intervention provides a prompt and satisfactory resolution of the problem⁸. In this case, the surgical procedure was postponed since the tumefaction of the tongue was remarkably reduced by needle aspiration of the lesion and the child had no breathing or feeding difficulties. However, the cyst gradually enlarged. The surgical resection of the complete lesion was performed when the patient was two months old. There has been no evidence of recurrence during the 22-months follow-up. A timely prenatal diagnosis is fundamental. Most congenital masses are corrected by postnatal surgery⁴. The possibility of performing an EXIT procedure should be considered in those cases of suspected obstruction of the upper airways. Tongue mucocele must be ruled out in the differential diagnosis of fetal oral cavity cystic lesions. We report the earliest diagnosis of tongue mucocele described in a prenatal ultrasound examination. Early clinical assessment, differential diagnosis and magnetic resonance imaging allow clinicians to diagnose and treat this rare condition with surgery in early infancy.

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