A Case of Imperforate Wharton Duct

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Congenital oral masses are rare entities. The establishment of formal fetal diagnostic teams has led to an increased antenatal detection of such lesions. The congenital ranula is a distinct entity from the more familiar variant presenting later in life. The congenital variant may result from an anomaly of the Wharton duct with subsequent dilation of the duct. The variant presenting later in life is the more familiar mucous extravasation phenomenon in the floor of the mouth. Management of the congenital ranula is distinct from its noncongenital counterpart and more conservative and is discussed in the present report.

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Congenital oral masses are rare entities. The establishment of formal fetal diagnostic teams has led to an increased antenatal detection of such lesions. The congenital ranula is a distinct entity from the more familiar variant presenting later in life. The congenital variant may result from an anomaly of the Wharton duct with subsequent dilation of the duct. Commonly presenting later in life is the more familiar mucous extravasation phenomenon in the floor of the mouth. Management of the congenital ranula is distinct from its noncongenital counterpart, is more conservative, and is discussed in the present report.[1-6]

Report of Case

A healthy 5-month-old female infant was referred to the University of Michigan Department of Pediatric Oral and Maxillofacial Surgery clinic on October 2011 for evaluation of bilateral cysts under the tongue first noted at birth. The infant was born uneventfully at 37 weeks after a pregnancy complicated only by preeclampsia. There was no airway obstruction at birth, but she had difficulty feeding. This was aggravated by reflux, constipation, and milk protein allergy. Soon after, she was readmitted for jaundice requiring 7 days of hospitalization. Her medical history was remarkable only for gastroesophageal reflux managed first with an H_2 receptor antagonist and then with a proton pump inhibitor. Her only medication at presentation was omeprazole. There was no history of surgery or trauma or a relevant family history. The parents denied any fevers, chills, nausea, vomiting, anorexia, or submandibular edema. She was found to have a milk allergy (unspecified protein allergy), with restoration of normal feeds with the use of hypoallergenic formula containing predigested amino acids. Otherwise, she was meeting all appropriate developmental milestones.

Physical examination showed a normal healthy-appearing infant without any distress. Vital signs were within normal limits. There were no palpable masses and no tenderness to palpation of the head and neck. Intraoral examination showed fullness in the floor of the mouth bilaterally, with a bluish hue along the path of the Wharton ducts, which was worse on the left. In addition, there was a small raised 0.5-cm submucosal lesion that appeared to represent an area of mucous extravasation located just medial to the expected opening of the left Wharton duct orifice. The floor of mouth was soft and compressible without tenderness. The presence of salivary drainage from Wharton duct orifices during digital submandibular gland stimulation could not be appreciated. A working differential diagnosis of bilateral congenital Wharton duct atresia, duplication of the submandibular ducts or accessory duct, foregut duplication cyst, and a lymphatic malformation was considered.[7-13] Other lesions arising in the floor of the mouth of an infant include epidermoid cyst, dermoid cyst, teratoma, hemangioma, and thyroglossal duct cyst.

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The congenital nature of the complaint did not favor a traditional ranula as the diagnosis.

An examination under anesthesia was arranged to permit probing of the ducts and, if possible, obtain a computed tomographic (CT) sialogram. The ducts could not be cannulated, and saliva could not be expressed from either duct. The CT scan depicted mild to moderate sialectasis of the left Wharton duct with less distinct enlargement on the right. No other lesions were noted and the submandibular and sublingual glands were intact. A working diagnosis of bilateral congenital Wharton duct orifice atresia was made. Reported treatment options include observation, puncture of the ducts, duct puncture with stenting, or sialodochoplasty with marsupialization. After discussion of the risks, benefits, and alternatives, the parents opted for the latter.

On the day of surgery, the patient was 7 months old and weighed 7.3 kg. Under general anesthesia with a nasoendotracheal tube, the region of the floor of the mouth was inspected clinically and sonographically. A small hockey-stick transducer probe at a frequency of 9 Hz was used to visualize the dilated Wharton ducts longitudinally and confirm the diagnosis. The ducts were hypoechogenic and compressible and greater at the left than at the right. Attention was directed to the left side, where a 1.5-cm incision was carried through the mucosa and submucosa with a fine-tip electrocautery in pure cut mode. A blunt and sharp dissection was used with a small Metzenbaum scissor to isolate the Wharton duct. The duct appeared significantly dilated. A small incision was made anteriorly and lacrimal probes were used to cannulate the duct (Figs 1, 2). A 1-cm incision was made at the superior aspect of the duct over the lacrimal probe. The duct was spatulated and marsupialized by suturing the exposed duct epithelium to surrounding oral mucosal epithelium with 7-0 polyglactin (coated Vicryl Plus Antibacterial suture, Ethicon, St. Louis, Missouri) sutures on a side-cutting needle. The patency of the duct was confirmed at the completion of marsupialization (Fig 3). The same procedure was performed on the right side. There was only minimal bleeding and no intraoperative complications were encountered. The patient was kept overnight for observation and intravenous fluids owing to concerns about postoperative oral intake.

At the 3-month follow-up visit and at 10 months of age, there were no episodes of recurrent floor-of-mouth edema. The submandibular glands were soft and nontender, with unhindered salivary flow from the Wharton ducts.

Discussion

A congenital anomaly refers to an abnormal condition present at birth. Etiologic factors may include chromosomal abnormalities, errors of morphogenesis (ie, neural crest cell migration), intrauterine environment, and maternal exposures. Congenital anomalies are generally classified as malformations, deformities, and disruptions. An imperforate submandibular duct may be a malformation resulting from an intrinsically abnormal developmental process in embryogenesis (ie, failure of apoptosis and canalization). Understanding these events in fetal development will guide the astute clinician in developing a diagnosis based on prenatal ultrasound or postnatal examination findings. This is contrast to developmental (from postuterine life on) or acquired (from trauma or pathology) anomalies, which have different etiologies and may involve different treatment modalities.

During the 10th and 12th weeks of gestation, the submandibular gland begins to form as an epithelial outgrowth surrounded by the mesenchyme forming the floor of the mouth. Differentiation of this ectomesenchyme gives rise to acinar and ductal cells. From 18 to 25 weeks, the gland acquires a connective tissue capsule and there is rapid proliferation leading to a steep increase in the number of gland lobules and canalized tubules. Interlobular ducts start to appear at 22 weeks. By 28 weeks, 95% of tubules are canalized. Distal perforation of the Wharton duct into the medial paralingual sulcus is believed to be one of the last processes in its development.[14]

Congenital atresia of the Wharton duct occurs when solid cord cells fail to undergo apoptosis and thereby impede canalization of the duct into the oral cavity. It is a rare anomaly that presents with sialectasis or dilation of the Wharton duct. This results from accumulation of saliva that distends the duct, which can be seen as an elongated, cystic, compressible, bluish mass in the floor of the mouth. The greatest area of fluctuance would be expected along the plica sublingualis, which corresponds to the duct's trajectory.[14]

This entity must be included in the differential diagnosis of any fluid-filled lesion of the oral cavity that may be diagnosed at prenatal imaging. Compared with solid lesions, such as granular cell tumors or teratomas, the echogenicity and signal characteristics of these lesions are quite different when imaged with fetal ultrasound or magnetic resonance imaging (MRI). In addition, Doppler ultrasound can be used to differentiate these from vascular lesions. For lesions likely to cause airway obstruction, plans can be made for an ex utero intrapartum treatment procedure with intubation, tracheostomy, or extracorporal membrane oxygenation. After pregnancy, emergency treatment of airway obstruction by any fluid-filled sublingual lesion also might include needle decompression and aspiration.[15]

In a case report by Kawahara et al,[16] the investigators found 13 cases of congenital dilatation of the