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Review Article

Sublingual cysts of different entities in an infant – A case report and literature review

Jan Oliver Voss[a,*], Salvatore Buehling[a], Nadine Thieme[b], Christian Doll[a], Kathrin Hauptmann[c], Max Heiland[a], Nicolai Adolphs[a], Jan-Dirk Raguse[a]

[a] Charité – Universitätsmedizin Berlin, Corporate Member of Freie Universität Berlin, Humboldt-Universität zu Berlin, Berlin Institute of Health, Department of Oral and Maxillofacial Surgery, Augustenburger Platz 1, 13353, Berlin, Germany
[b] Charité – Universitätsmedizin Berlin, Corporate Member of Freie Universität Berlin, Humboldt-Universität zu Berlin, Berlin Institute of Health, Diagnostic and Interventional Radiology and Nuclear Medicine, Augustenburger Platz 1, 13353, Berlin, Germany
[c] Charité – Universitätsmedizin Berlin, Corporate Member of Freie Universität Berlin, Humboldt-Universität zu Berlin, Berlin Institute of Health, Institute of Pathology, Campus Mitte, Charitéplatz 1, 10117, Berlin, Germany

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ABSTRACT

Background: Congenital cysts and fistulas of the neck are common in children, often located in the head and neck area. Belonging to the group of tumor-like conditions, dermoid and epidermoid cysts are dysontogenetic lesions with seldom multiple co-occurrences in infants.
Case report: We report on a nine-month-old female with a persisting congenital fistula of the tongue. The patient was admitted with acute poor feeding and hypersalivation, which started within the last 24 h. Magnetic resonance imaging detected a fistula of the tongue connected to sublingual cystic lesions. Intraoral surgical removal of three cystic lesions and the fistula was performed under general anesthesia. Histopathological analysis confirmed the coexistence of an epidermoid cyst and two dermoid cysts.
Conclusion: Sudden feeding difficulties in combination with dysphagia and tongue displacement in pediatric patients pose an emergency situation that requires prompt diagnostic clarification. A persisting congenital fistula of the tongue is a clear indication of dysontogenetic lesions, including malformations, tumors, and tumor-like lesions. Congenital sublingual cysts are rare in infants, but can be life threatening when present. Surgical excision with histopathological analysis is essential to exclude any form of malignancy and malignant transformation.

1. Introduction

Epidermoid and dermoid cysts of the head and neck area are common, especially in pediatric service [1,2]. While most cysts in children are located in the neck, lesions located in the floor of the mouth are considered rare [2,3], especially those of congenital nature [4,5].

Epidermoid and dermoid cysts are considered to be dysembryogenetic lesions derived from enclavement of ectoderm, developing during the third and fourth week in utero during midline closure of the bilateral first and second branchial arches [6]. Based on histopathological analysis, dysontogenetic cysts of the floor of the mouth can be divided into a) epidermoid, b) dermoid, and c) teratoid cysts [7]. Cysts originate during embryological development when pharyngeal arch growth takes place; therefore derivatives of the three germ layers may be found [7].

Clinically, different anatomical locations should be distinguished in relation to the geniohyoid muscle: a sublingual position (above) and a submental position (below). While sublingual cysts lead to displacement of the tongue with swelling impairment, submental cysts extend from the mandible to the hyoid bone giving the impression of a “double chin” appearance [6,7]. Furthermore, a differentiation between lateral and medial cyst location can be made [6]. Even though epidermoid and dermoid cysts are regarded as congenital pathologies, clinical manifestation may be uneventful in the beginning. Most lesions become clinically evident from 15 to 35 years of age [5,7].

* Corresponding author. Department of Oral and Maxillofacial Surgery, Charité – Universitätsmedizin Berlin, Campus Virchow-Klinikum, Augustenburger Platz 1, 13353, Berlin, Germany.
E-mail addresses: jan.voss@charite.de (J.O. Voss), salvatore.buehling@googlemail.com (S. Buehling), nadine.thieme@charite.de (N. Thieme), christian.doll@charite.de (C. Doll), kathrin.hauptmann@charite.de (K. Hauptmann), max.heiland@charite.de (M. Heiland), nicolai.adolphs@charite.de (N. Adolphs), jan-dirk.raguse@charite.de (J.-D. Raguse).

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Although these cysts are initially painless [5], infection of cystic lumen might lead to pain and discomfort, as well as progressive swelling [8]. Depending on cyst size, there is a wide range of clinical symptoms, such as respiratory problems, dysphagia, and difficulties in speaking and/or drinking [5]. In the case of increasing tumor size, displacement of the tongue is seldom apparent with life threatening consequences [9,10]. Clinical examination is followed by morphological imaging. Depending on the age of the patient and the localization of the cystic lesion, ultrasound is frequently used [2,11]. However, magnetic resonance imaging is another option, especially in cases with fistula and multilocular expansion. Complete exstirpation/surgical excision of the cyst is the treatment of choice.

Here, we present a rare case of the coexistence of an epidermoid and two dermoid cysts with a persisting fistula of the tongue in an infant.

1.1. Case report

A 9-month old female patient was admitted to the pediatric emergency department with fever (38.4 °C) and a new sucking weakness resulting in poor feeding due to a progressive sublingual swelling with increased salivation, which had started within the last 24 h. According to the parents, the infant had a postnatal diagnosis of persisting fistula, which was regularly checked by a physician in private practice. However, the parents reported a yellowish liquid, which had drained from the cyst for the first time some days prior. Pre-medications and previous operations were negated.

Extraoral physical examination was uneventful. However, intraoral examination revealed a sublingual swelling/mass with an elevated and dorsally displaced tongue. Bimanual palpation of the submandibular gland was clinically unremarkable. A fistula of the tongue was not detectable during initial examination due to the displacement of the tongue and the strong non-compliance of the patient.

Magnetic resonance imaging (MRI) was performed in narcosis and revealed three sublingual cystic lesions plus one at the dorsal part of the tongue. T2 weighted images with and without fat saturation displayed inhomogenous, peripheral slightly hyperintense lesions with central parts of hypointensity, while on T1 weighted images, these cystic lesions showed a hypointense signal, measuring 20 × 20 × 10 mm (individual cavity dimensions: cranial: 10 × 9 × 9 mm; caudal: 20 × 9 × 11 mm). Cystic lesions were located sublingually between the genioglossus muscles and cranial to the mylohyoid muscle (Figs. 1–3). In addition, a fistula about 4 mm in width was detected starting from the cranial cystic lesion and ending at the midline anterior part of the tongue (Fig. 2).

Diffusion weighted imaging (DWI) demonstrated low apparent diffusion coefficient (ADC) values, corresponding to a diffusion restriction compatible with epidermoid cysts, dermoid cysts or abscess formations (Fig. 4).

Additionally, at the dorsal edge of the tongue, an extremely sharp contoured, in T2w images strongly hyperintense lesion measuring 5 × 5 × 5 mm was detectable, with strong hypointensity in T1w images, no enhancement of contrast agent and with high ADC values corresponding to released diffusion without uptake of contrast agent (Figs. 2–4).

Blood analysis revealed a C-Reactive-Protein (CRP) level of 5.0 mg/l with normal blood count. However, intravenous antibacterial treatment with Cefuroxim adapted to body weight was initiated. Still under general anesthesia, the patient was taken directly to the surgery theater for further exploration and surgical treatment. On palpation, a yellowish fluid was drained from a fistula on the back of the tongue. The tongue was pulled upward by two traction sutures. Blunt probing of the fistula revealed a deep extension of the fistula into the floor of the mouth (Fig. 5).

By circular incision, the fistula was removed from the tongue. In total, three connected cystic lesions were excised in toto by an incision parallel to the frenulum of the tongue followed by histological evaluation (Figs. 6 and 7). Lastly, the retention cyst at the dorsal edge of the tongue was punctured without further histopathological analysis. Due to the swelling of the floor of the mouth, the patient was taken to the postoperative care unit for further monitoring after surgery. On the first postoperative day, the patient was extubated.

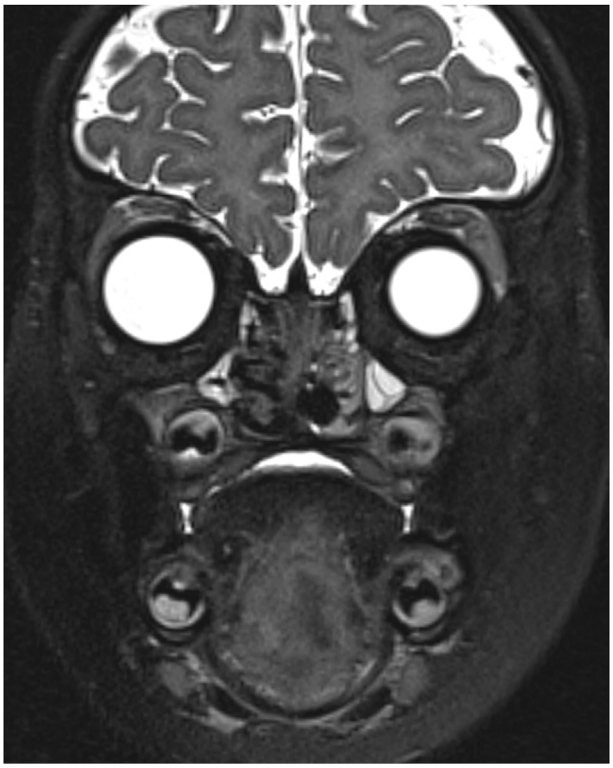

Fig. 1. Diagnostic imaging. T2wTSE sequence with fat saturation with intravenous contrast agent in coronal plane demonstrating the sublingual altered cystic lesion cranial to the mylohyoid muscle.

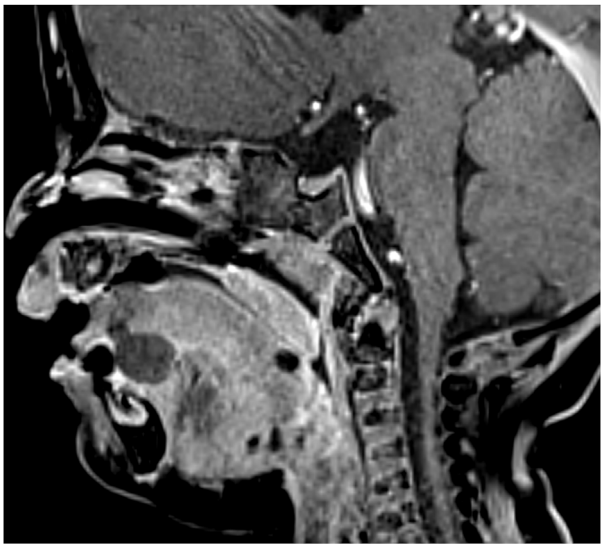

Fig. 2. Diagnostic imaging. T1w GE sequence with intravenous contrast agent in sagittal plane demonstrating the sublingual altered cystic lesion cranial to the mylohyoid muscle and a cystic lesion located submucosally in the dorsal third of the tongue. In this Figure, the fistula ends at the anterior part of the tongue.

Histopathological examination revealed a ruptured epidermal cyst (Fig. 8), as well two dermoid cysts (Fig. 9, representative for both cysts) with severe recurrent and partly absceding inflammation infiltrating the local environment. A malignancy of the lesion was excluded.