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Brief Report

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# A Rare Submandibular Space Foregut Duplication Cyst Mimicking a Lymphatic Malformation in a Child

**Teresa Gross Kelly*, Subramanian, David C Moe, Robert H. Chun, Paula E. North, Mia S. Kelly and David C. Howlett**

[1]Departments of Radiology Children's Hospital and Health System of Wisconsin and Medical College of Wisconsin

[2]Department of Otolaryngology, Children's Hospital and Health System of Wisconsin and Medical College of Wisconsin

[3]Department of Pathology, Children's Hospital and Health System of Wisconsin and Medical College of Wisconsin Milwaukee, WI, USA

[4]Department of Radiology, Eastbourne District General Hospital East Sussex Hospitals NHS Trust Eastbourne, UK

[5]Department of Radiology, Children's Hospital of Pittsburgh Pittsburgh, PA, USA

***Corresponding author:**

Teresa Gross Kelly,
Departments of Radiology Children's Hospital and Health System of Wisconsin and Medical College of Wisconsin, USA, Tel: 414-266-1686; Fax: 414-266-1525, Email: tkelly@chw.org

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## 1. Introduction

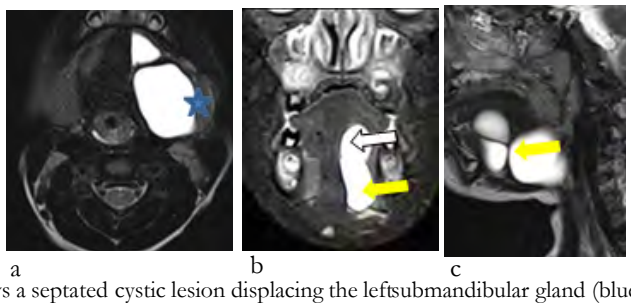
A submandibular space (SMS) bronchopulmonary foregut cystic malformation (BPFCM) was mistaken for a lymphatic malformation (LM) and treated unsuccessfully with sclerotherapy. The purpose of this article is to describe the imaging features of this rare SMS BPFCM so that it can be differentiated from LMs and treated appropriately. We propose that BPFCM in the submandibular space should be included in the list of differential diagnostic possibilities when encountering a septated submandibular cystic mass.

## 2. Case Report

A 3½ year old girl presented with a recurrent lump beneath the left jaw. On physical examination beneath the angle of the left mandible, a soft, nontender mass, with no overlying erythema or induration, was palpated. The floor of the mouth appeared normal and she was afebrile. She reportedly had swelling in the same location when she was 1 year old. At that time, an ultrasound revealed a cystic mass. Surgical excision was recommended, but the mass spontaneously resolved. When the mass recurred, MRI was performed revealing a septated cystic mass located medial to the left submandibular gland spanning the left sublingual and submandibular spaces (Figure 1a). No solid component or restricted diffusion was observed. Mild septal and peripheral wall enhancement was noted.

As lymphatic malformations (LM) are transpatial multicystic masses, a LM was considered to be the most likely etiology. The differential diagnosis included a plunging ranula or a second branchial cleft cyst (BCC). However, BCCs are typically situated posterior to the submandibular gland rather than medial to it, making BCC less likely. A ranula was also not as likely because a mass could not be palpated along the floor of the mouth, which is characteristic for this entity. Based on a presumptive diagnosis of a LM, treatment with ultrasound-guided sclerotherapy was initiated (Figure 2 a). The patient underwent a total of three rounds of sclerotherapy at three-month intervals. However, in spite of multiple treatment attempts, there was no progress and the mass continued to enlarge. Therefore, a CT scan was ordered for further evaluation. The CT scan showed a cyst surrounded by a thickened wall containing scattered coarse calcifications (Figure 3). It was postulated that the calcifications could be related to the multiple sclerotherapy attempts. However, upon further review of the prior limited ultrasound images obtained during sclerotherapy, subtle echogenic foci in the wall of the lesion could be detected (Figure 1, 2b). The lesion was mulitseptated and located well away from the midline, spanning the left submandibular and sublingual spaces. No additional sclerotherapy attempts were made and the patient underwent surgical resection, revealing a dumbbell

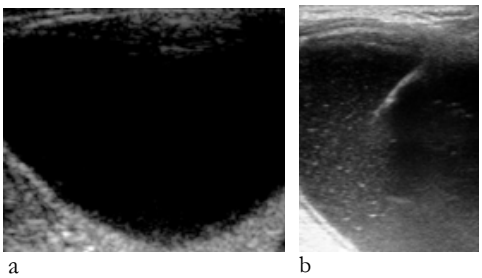
shaped mass (Figure 4). Histologic analysis exhibited elements of both ventral foregut differentiation (pseudostratified columnar epithelium) and dorsal foregut differentiation (pancreatic tissue and gastric epithelium) (Figure 5). The presence of pseudostratified ciliated respiratory epithelium, pancreatic and gastric type epithelium in the patient's excised left submandibular cystic mass was compatible with a bronchopulmonary foregut cystic malformation (BPFCM). There was no histological evidence of a lymphatic malformation.



a b c

(a) Axial FS T2 weighted image shows a septated cystic lesion displacing the leftsubmandibular gland (blue star) laterally. (b) STIR coronal images shows the cystic lesion traversing the sublingual space (white arrow) and submandibular space (yellow arrow). (c) STIR coronal images show the cystic lesion traversing the submandibular space (yellow arrow).

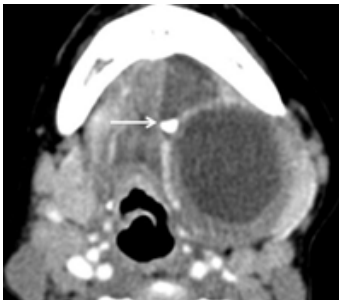
**Figure 1**: MRI reveals a septated submandibular cystic mass thought to represent alymphatic malformation, less likely a plunging ranula.



a b

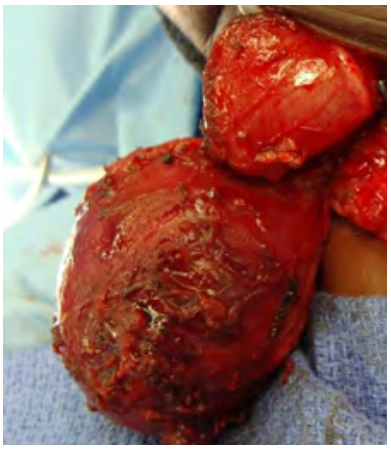
(a) Pre-sclerotherapy diagnostic ultrasound shows a multiseptated cystic mass containing internal echos and exhibiting enhanced through transmission. (b) Usingultrasound guidance, approximately 20 ccs of cloudy white fluid were aspirated and Picibanil (OK432) was injected into the cystic lesion.

**Figure 2**: Ultrasound guided sclerotherapy was performed.



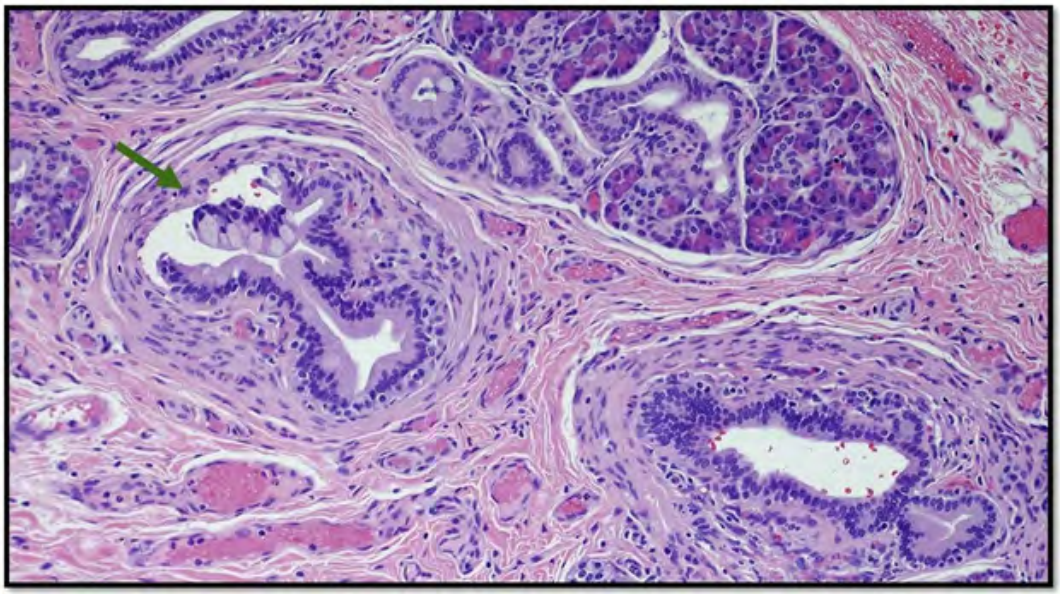
(a) This CT shows interval enlargement of the cyst and a coarse calcifications in asurrounding thickened wall (white arrow).

**Figure 3**: Unenhanced axial CT image was acquired after multiple sclerotherapyattempts were made.



(a) A bilobed dumbbell shaped mass was excised without complications.

**Figure 4**: Gross surgical specimen.



(a) High power view (original magnification 200x) showing foci of pancreatic tissue (top) and gastric foveolar epithelium containing isolated goblet cells (arrow).

**Figure 5**: Pathologic histological analysis revealed a bronchopulmonary foregutcystic malformation.

## 3. Discussion

In the developing embryo, the dorsal foregut gives rise to the upper gastrointestinal tract whereas the ventral foregut develops into the lower respiratory tract [1]. Abnormal budding from the foregut gives rise to three types of foregut malformations: (a) the first type is due to abnormal budding from the dorsal foregut, giving rise to an enteric duplication cyst which can be found anywhere along the alimentary tract from the oral cavity to the rectum, (b) the second type results from abnormal budding from the ventral foregut resulting in a bronchogenic cyst, (c) and the third type represents an accessory lung bud that gives rise to a rare type of cyst comprised of both dorsal and ventral foregut components called a bronchopulmonary foregut cystic malformation (BPFCM) [1]. The submandibular space cystic mass mistaken for a LM was the third type, a BPFCM, as it contained elements of both ventral foregut differentiation (pseudo-stratified ciliated respiratory epithelium) and dorsal foregut differentiation (pancreatic tissue and gastric-type epithelium). Cyst wall calcifications seen in this case have also been reported in bronchogenic cysts and in branchial cleft cysts but they are not a common finding in lymphatic malformations. Therefore, the presence of cyst wall calcifications could help to differentiate LMs from either BPCFM or BCCs when encountering a multicystic mass in the submandibular space. The absence of restricted diffusion argued against the possibility of a dermoid/epidermoid cyst. The inability to palpate a mass along the floor of the mouth was not compatible with a ranula, and finally, the location of the mass was not compatible with a second branchial cleft cyst, commonly located between the sternocleidomastoid muscle and submandibular gland rather than medial to it, as in this case. In general, head and neck foregut duplication cysts are not common. Most foregut duplication cysts occur in the chest and abdomen [2]. In one retrospective review of 23 head and neck foregut duplication cysts, most (52%) foregut duplication cysts were found in the oral cavity, 5% in the anterior tongue, and 7% in the floor of mouth [2,3]. BPFCMs specifically in the submandibular space without involving the floor of mouth were not specifically described. If left untreated, foregut cystic malformation complications can arise, including peptic ulceration, bleeding, perforation, as well as sinus tract formation exuding chronic mucous sections [2,4]. In order to prevent such complications and to provide the appropriate treatment, accurate diagnosis of foregut cystic malformations in the submandibular space is crucial [5-7].

Based upon the imaging findings presented in this case, we propose that the presence of wall calcifications can help differentiate a LM from a BPFCM. The absence of restricted diffusion can distinguish a foregut duplication cyst from a dermoid cyst. Bronchopulmonary foregut cystic malformations should be remembered as a diagnostic possibility when encountering a transpatial, septated cystic lesion located in the pediatric submandibular space.

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