Case Report

Massive Sublingual Bronchogenic Cyst with Airway Compromise

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Abstract

Bronchogenic cysts are primary cysts of the mediastinum. In rare situations, it can present in the oral cavity as a swelling of the tongue and the floor of the mouth. Usually, it is not a life-threatening clinical entity. In this report, we present a massive sublingual bronchogenic cyst with life-threatening clinical characteristics in an adult patient of a Nigerian tertiary-level medical center and the management of the case.

Keywords: Cavity, cysts, foregut

Introduction

Lingual cysts are uncommon clinical conditions which are known to arise from the epithelial lining of the gut or respiratory system.[1] Embryonal residues of the gut or respiratory epithelium, captured in the tongue which subsequently underwent cystic degeneration have been reported in the literature.[1-3] These epithelial remnants have been described as lingual bronchogenic cysts (or simply bronchogenic cysts) when derived from the respiratory epithelium and it is considered by some authors to be choristomas, heterotopic cysts, or foregut duplication cysts.[2-5]

Bronchogenic cysts are the most common primary cysts of the mediastinum, and by virtue of this description, they are located in the thoracic cavity.[2] In rare situations, this cyst is found in the extrathoracic location, the most commonly reported site being the tongue.[6] Lingual bronchogenic cyst can present on the tip of the tongue,[6] the ventral surface of the tongue,[2] the dorsal surface of the tongue,[7] and the substance of the tongue as macroglossia[3] or sublingual (floor of the mouth).[8] Although it is more commonly found in male and mostly reported in children, this congenital oral cyst has been reported in adults.[8,9]

This article describes a case of the massive life-threatening sublingual cyst with respiratory epithelium in our hospital and highlights of our management of the patient.

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Case Report

A 27-year-old male fashion designer presented at the dental clinic of the Federal Medical Centre, Ebute-Metta, Lagos, Nigeria, on account of worsening airway obstruction, difficulty in swallowing, and speech impairment due to a massive swelling in the substance of the tongue and floor of the mouth. The swelling was noticed shortly after birth, according to the mother. It increased in size and was sometimes noticed to recede. Three months before presentation, the patient accidentally bit the swelling with the consequent rapid increase in size. There was associated pain which disturbs sleep, but there was no history of bleeding or discharge. The increase in the size of the swelling and pain affected feeding with resultant progressive weight loss and generalised body weakness. There was no history of fever, night sweats, or similar swelling in other parts of the body. An episode of fainting was reported about 10 h before presentation, but the patient regained consciousness within a few minutes. This necessitated his presentation at our facility.

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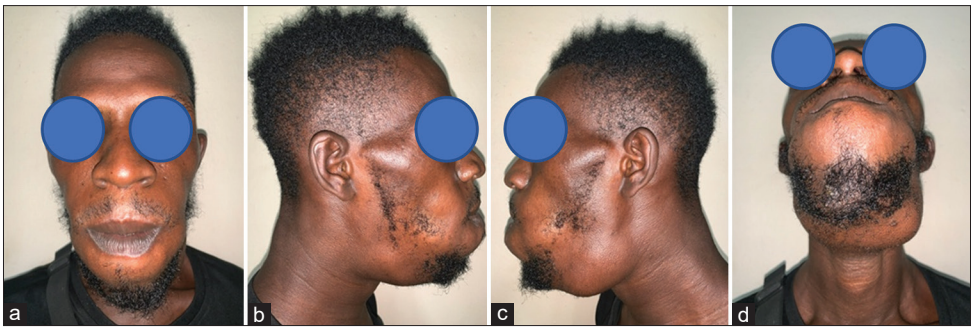

Figure 1: (a-d) Clinical photograph of patient at presentation

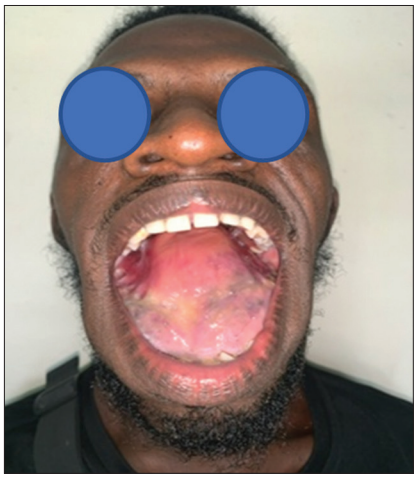

Figure 2: Intraoral photograph of patient at presentation

Examination revealed an emaciated young-looking man with bulging eyeballs, flaring of the nasal ala, and sunken cheeks giving a gaunt face. The patient was not pale, acyanotic, and not jaundiced. There was an increase in facial height with class III facial profile. No abnormality of the temporomandibular joints was observed. There was a diffuse but firm swelling involving the submental and submandibular region, which appeared to be extending to the upper part of the neck bilaterally and appeared to be more pronounced on the left. The swelling is 14 cm × 12 cm in dimension. Level IA and B lymph nodes could not be examined due to the swelling. Level II, III, and IV lymph nodes were not enlarged [Figure 1].

Intraorally, there was dental anarchy evidenced by flaring and proclination of the lower anterior teeth. There was compensatory proclination of the upper anterior teeth giving a bimaxillary prognathic facie. Patient's oral hygiene was poor evidenced by gross plaque and calculus accumulation. The lower anterior, canine, and premolars were mobile. Intraoral swelling extended over the floor of the mouth into the substance of the tongue. There was no observable demarcation of the floor of the mouth and the tongue, and there was total obliteration of the alveololingual sulcus. The swelling was, however, firm and not tender [Figure 2].

An initial clinical assessment of lymphangioma was made. Craniocervical magnetic resonance imaging (MRI) and anterior neck ultrasonography (USS) were requested.

The USS showed a thick-walled hypoechoic cystic mass with heterogenous mobile echogenic floaters extending from the right jaw angle to the left jaw angle through the submandibular space. No solid component was seen, but the cystic mass has an estimated volume of 450 cm^3, 7 mm away from the skin, and a thickness of 10 cm in the submandibular region. The MRI [Figure 3] shows a large expansile thick-walled cystic mass with a lobulated outline occupying the sublingual space and extending to fill the oral cavity, compressing the lingual soft tissues as well as the oropharynx. The mass is homogenously hyperintense on T2-weighted images.

An assessment of impending airway obstruction and difficult intubation necessitated a plan for marsupialisation under general anesthesia through elective tracheostomy. The cyst contained milky white fluid with cheesy particles [Figure 4]. Sterile gauze plug was inserted into the marsupialised cavity and changed daily. Tracheostomy care and decannulation were instituted over three postoperative days, and the patient was discharged home on antibiotics and analgesics after five days postoperative period. Insertion of sterile gauze at meal times and during sleep formed part of the home instructions that was given the patient.

Cyst excision under general anesthesia through nasotracheal intubation was carried out two months after the first stage. All hematological parameters, blood chemistry, and clotting profile were within the normal interval before surgery. A pack of gauze was tucked into the marsupialised cavity, and an incision was made along the cuff of the cavity. Blunt dissection was done following the thick cyst wall till the entire cyst capsule was eradicated en masse [Figure 5]. This was followed by layered and mucosal closure using 3/0 vicryl.

The patient recuperated rapidly on a daily dose of 1 g intravenous ceftriaxone and 75 mg of intramuscular diclofenac and was subsequently discharged home on oral antibiotics and analgesics on the fifth postoperative day.

Discussion

During the differentiation of the foregut in the third week of embryonic development, the foregut develops in two parts; a ventral part forms the respiratory tract and the dorsal part forms the gastrointestinal tract.[4] The anterior tongue arises

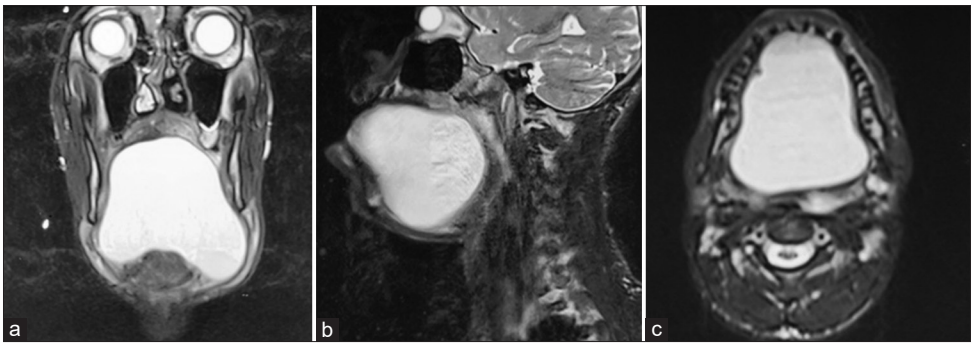

Figure 3: (a-c) Magnetic resonance imaging (MRI) of the lesion

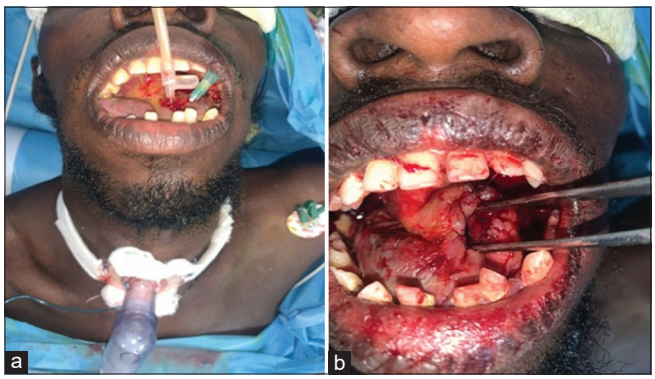

Figure 4: (a) Tracheostomy tube in-situ, (b) Marsupialisation

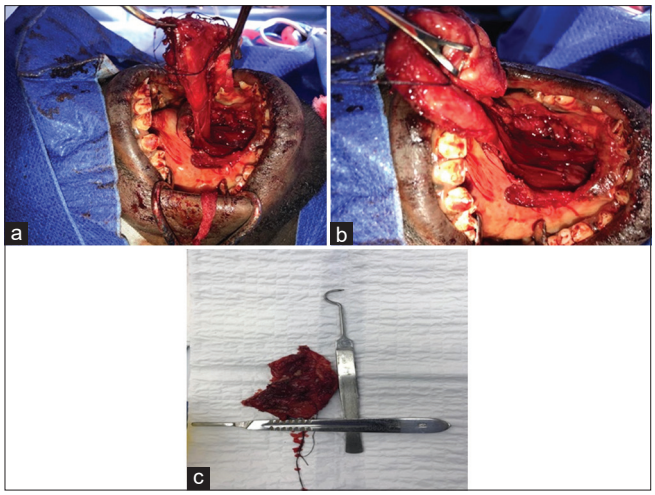

Figure 5: (a & b) Clinical photograph of cyst excision, (c) Surgical specimen (cyst)

from the tuberculum impar and the adjacent regions of the mandibular arches, while the posterior portion is from the ventromedial ends of the second branchial arches. These paired portions fuse anterior to the hypobranchial eminence to form the copula with a later contribution of the mesoderm.[4] Owing to the proximity of the foregut and the developing branchial arches, a cyst arising from undifferentiated embryonic rest of the primitive gut may be trapped or misplaced in any part of the developing tongue or other parts of the oral cavity.[5] These could differentiate into gastrointestinal or respiratory epithelium depending on environmental and inductive influences.[5] Another theory suggests that bronchogenic cyst derives from multipotent stem cells which are capable of multidirectional differentiation. These cells proliferate and subsequently undergo cystic degeneration.

Irrespective of developmental origin, the most common sites for bronchogenic cysts are the mediastinum and the lung. These account for 75% and 25% of cases, respectively. The head-and-neck presentations occur in <1% of cases.[10] Most cases are discovered at birth but may appear later in life. Bronchogenic cysts can present as a completely asymptomatic or as a life-threatening condition and has a male predilection.[1,10] The clinical features are not distinctive from many other soft-tissue cysts of the tongue and floor of the mouth. MRI is the imaging of choice for suspected soft-tissue cysts, as in the case, but a definite diagnosis depends on histopathologic evaluation. When located in the sublingual area, the differentials may include lymphangioma, lymphoepithelial cyst, dermoid cyst, epidermoid cyst, salivary retention cyst, congenital cystic choristoma, and hemangioma. While histopathologic examination may easily distinguish bronchogenic cysts from endothelial cell-lined lesions such as lymphangioma, the distinction between bronchogenic cysts, esophageal cysts, and enteric cysts may be difficult due to their close embryologic relationship.[8]

This is the first case of bronchogenic cyst of the head-and-neck region to be reported in our facility. As reported in the literature, the authors had a diagnostic dilemma in view of the size, location, cheesy content of the cystic lesion and its appearance on the MRI. It is reported that bronchogenic cyst usually measures <3 cm but could measure up to 9 cm in diameter.[3] However, this index case measured 14 cm × 12 cm at presentation with signs of respiratory compromise. This may have been due to delayed presentation. The probable site of embryological rest entrapment was unclear as the lesion dissects into the tissue spaces of the tongue and transmylohyoid muscle into the upper part of the neck. By virtue of its location and clinical appearance, lymphangioma was a top differential.

Figure 6 shows that the lingual cyst lined by respiratory epithelium characterises a pathology distinguished histologically by well-differentiated ciliated, pseudostratified, columnar

epithelium overlying a fibrous connective tissue. This type of cyst may be diagnosed solely on the evidence of its morphology following previous recommendations.[1]

While surgical excision is the preferred method of treatment in the literature, the authors adopted marsupialisation (Partsch I procedure) and subsequent cyst excision (modified Partsch II procedure).[11] Marsupialisation with a surgical airway was done to relieve intraluminal pressure,[12] relieve the impending total airway obstruction, and assist shrinkage of the cyst.[12] Figure 7a and b show the clinical photographs of the patient one-day postmarsupialisation.

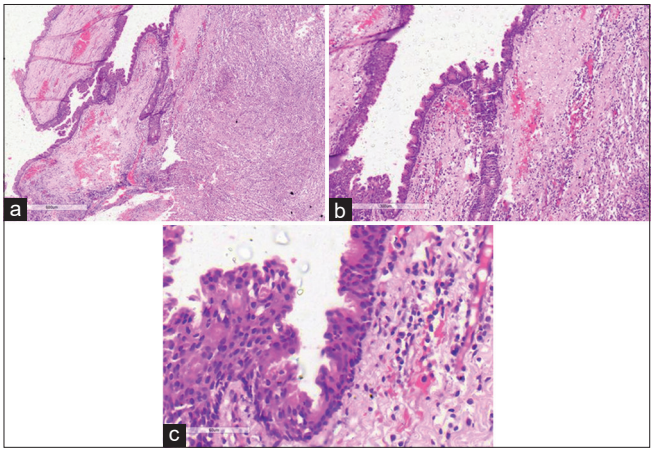

Figure 6: Micrograph at × 4 (a); ×10 (b); ×40 (c)

Figure 7: (a-b) Postoperative photograph one-day post-marsupialisation

This was followed by the excision of the cystic lining two months later. The two-stage approach was informed by the size of the lesion, the likelihood of iatrogenic injury to nerve supply of the tongue, the risk of inadvertent retention of cyst lining which could cause a recurrence, and postoperative airway compromise from postoperative edema. We prescribed preoperative IM dexamethasone 8 mg stat, 1 h before surgery, as well as perioperative antibiotics up to five days postoperative period. At six-month review [Figure 8], no evidence of recurrence was reported. The patient was counselled on the need for multidisciplinary care involving the periodontologist and orthodontist to restore normal dental alignment and overall oral health.

Conclusion

The rare occurrence of cystic degeneration of entrapped foregut epithelium in the oral cavity requires vigilance, especially because of the risk, it poses to the airway. This is more important in low-resource countries where patients do not have equitable access to advanced specialist care. Sound clinical evaluation supported by MRI is valuable but cannot be relied on for definitive diagnosis because of their similar presentations with lymphangioma and other developmental cysts. In large cases involving multiple structures of the head and neck, marsupialisation and later cyst enucleation/excision are the desired treatment option. With this approach, complete recovery is expected, and the long-term prognosis is excellent, with no recurrence reported. Our index patient is six months postintervention and no sign of recurrence or swelling has been noticed.

Compliance with ethical standard

Ethical approval (Approval Number: HREC 23-01) was obtained from the Ethical Board of the Federal Medical Centre, Lagos. Written informed consent was obtained from the patient for the use of his clinical history and photographs.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

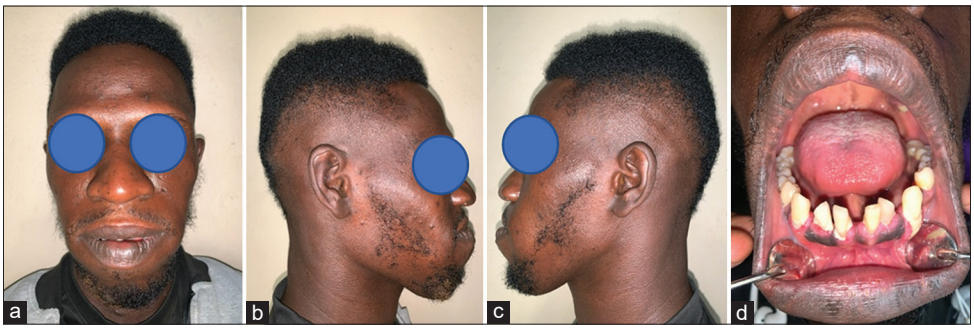

Figure 8: (a-d) Clinical photographs of patient six-month postoperative period

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Conflicts of interest
There are no conflicts of interest.

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