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Submental epidermoid cyst masquerading as a giant plunging ranula: a case report and review of literature

Eziagu UB[1*], *Inoh MI*[2]

[1]*Department of Histopathology, University of Uyo Teaching Hospital, Uyo, Akwa Ibom State, Nigeria.*
[2]*Department of Ear, Nose and Throat (ENT), University of Uyo Teaching Hospital, Uyo, Akwa Ibom State, Nigeria.*

Abstract

Submental epidermoid (epidermal) cyst is a very rare non-neoplastic cyst lined by squamous epithelium that arises from the floor of the mouth. It can sometimes clinically resemble a giant plunging ranula. This is a case report of a 20-year-old female who presented with a slow growing largely asymptomatic lower jaw swelling of 10 years' duration. A clinical diagnosis of giant plunging ranula was made. An excisional biopsy/corrective surgery was performed and subsequent histopathological evaluation revealed that this submental lesion was an epidermoid cyst. Thus, Submental epidermoid (epidermal) cyst can mimic a giant plunging ranula in its clinical presentation. Therefore, histopathologic evaluation is critical in making definitive diagnosis in such scenarios.

Key-words: Epidermal cyst, ranula, cysts, jaw, mouth

Introduction

Epidermoid (epidermal) cyst can be defined as a non-neoplastic cystic lesion that is lined by squamous epithelium and whose cystic content is keratin flakes, and can arise from any part of the body.[1–4] These cysts most commonly arise in the gonads (ovaries and testes) and rarely in the head and neck region; its occurrence in the oral cavity is indeed quite rare.[2–8] Aetiologically, these cysts can be classified as either congenital or acquired.[1–3]
Epidermoid cysts of the oral cavity usually start off as an asymptomatic lesion that slowly and progressively grows to a size that can elicit symptoms of varying severity proportionate to its size. These symptoms include difficulty in chewing, swallowing, breathing and talking.[3,5,8,9]

Corresponding Author: Dr. Eziagu, Uchechukwu Brian

Department of Histopathology,
University of Uyo Teaching Hospital, P.M.B. 1136,
Uyo, Akwa Ibom State, Nigeria. Postal code: 520001
Email: uchechukwu.eziagu@npmcn.edu.ng
Phone: +2348095794223

Sometimes, epidermoid cyst can indeed mimic a giant plunging ranula in the floor of the mouth in its clinical presentation, thus creating a scenario that increases the likelihood for its misdiagnosis.[10–12] In such scenarios, histopathological evaluation of this kind of masquerading lesion becomes the gold standard in arriving at a definitive diagnosis.[2,3,8,10–12]
The aim of this case report and literature review, therefore, is to show/explore the key role that histopathological evaluation of head and neck mass lesions (such as our index clinically diagnosed giant plunging ranula following a successful surgical excision) play in making accurate definitive head and neck histopathologic diagnoses (being submental epidermoid cyst, in this case, which was both rare and unsuspected) as well as ruling out close differentials/mimics. Furthermore, we aim to review relevant literature in order to characterise this lesion both pathologically and radiologically.

Case Report:

A 20-year-old female student of African descent who presented to our ear, nose and throat (ENT) outpatient clinic with complaint of 10 years' history

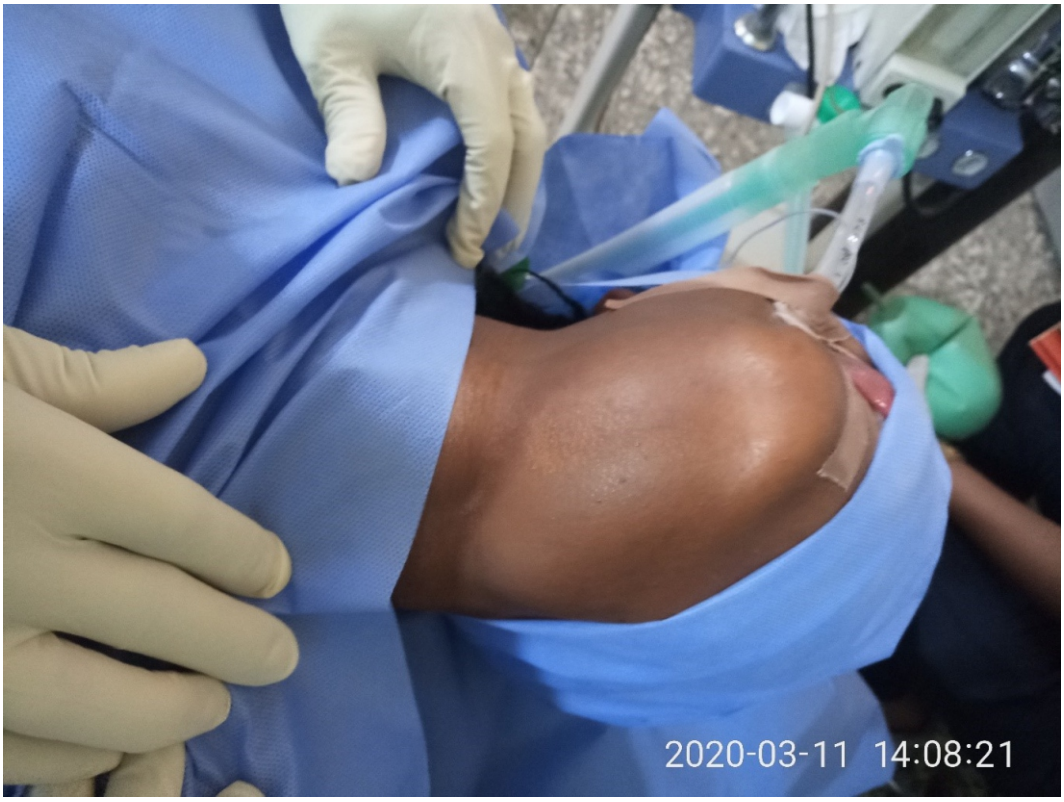

Figure 1: showing the huge submental mass in the theatre, just before the commencement of the corrective surgery. This mass neither moved with swallowing nor with the protrusion of the tongue. Note also that the skin over this submental mass appeared healthy with no scarification marks or engorged vessel.

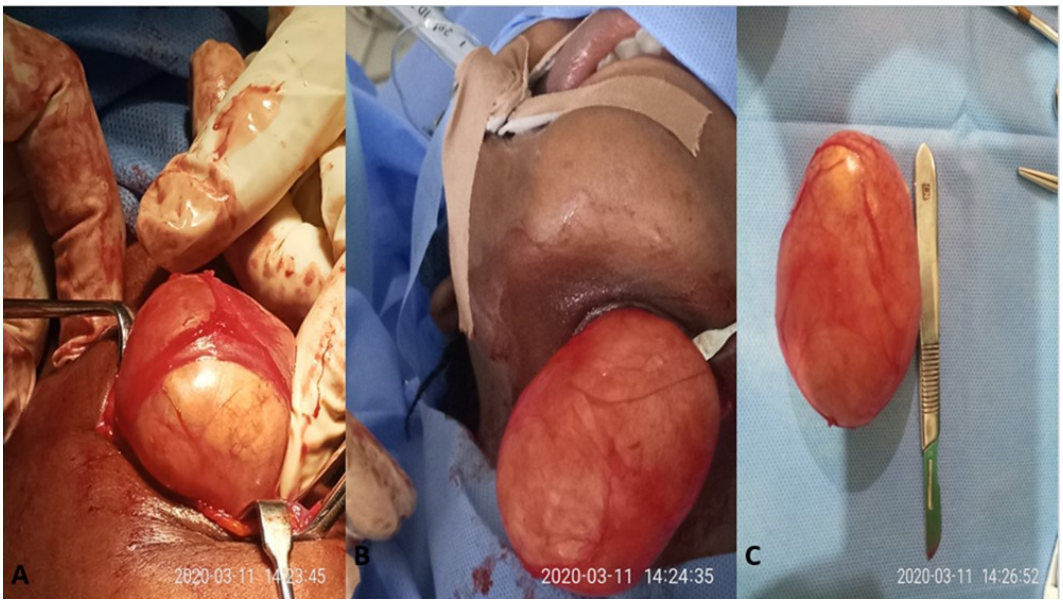

Figure 2 [A – C]: showing a huge oval well circumscribed cystic mass surgically excised from the submental region through a skin incision; this excised cystic mass lesion was soft, fluctuant, well circumscribed with fibrous attachment but no vascular attachment, while measuring 9.5cm in length, 5cm in width and 0.25kg in weight.

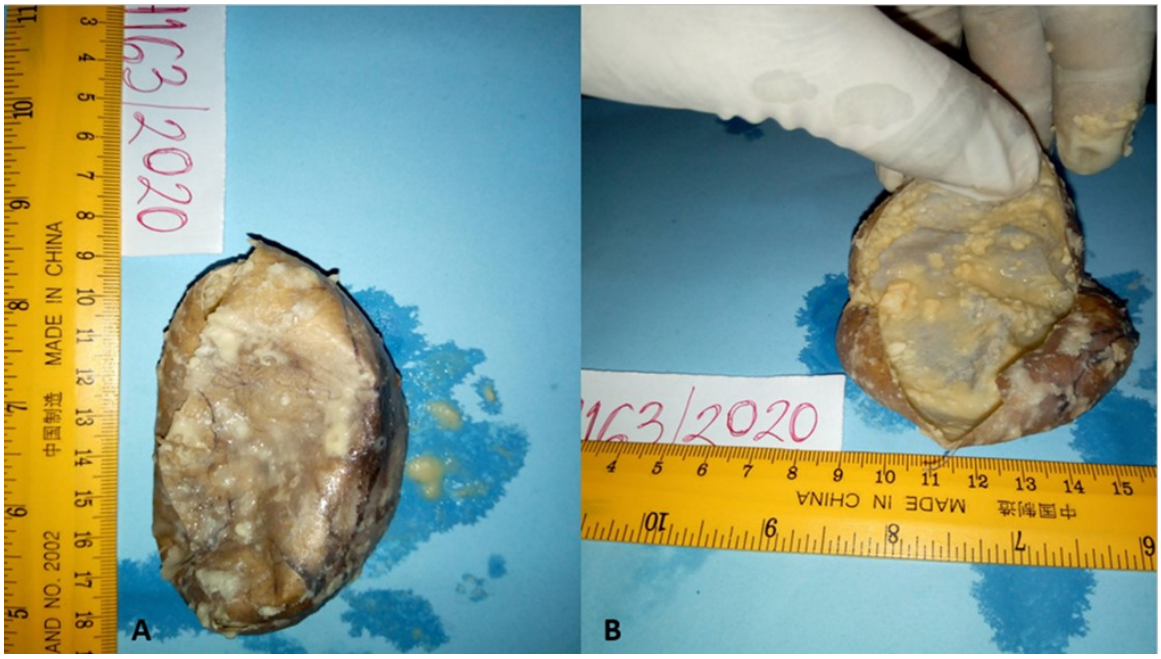

Figure 3 [A, B]: Gross photographs of the specimen following surgical cut-up; (**A**) shows the external surface of the collapsed cystic structure, (**B**) shows the inner surface of the cystic structure with remnant tan-grey cream coloured semi-solid substance.

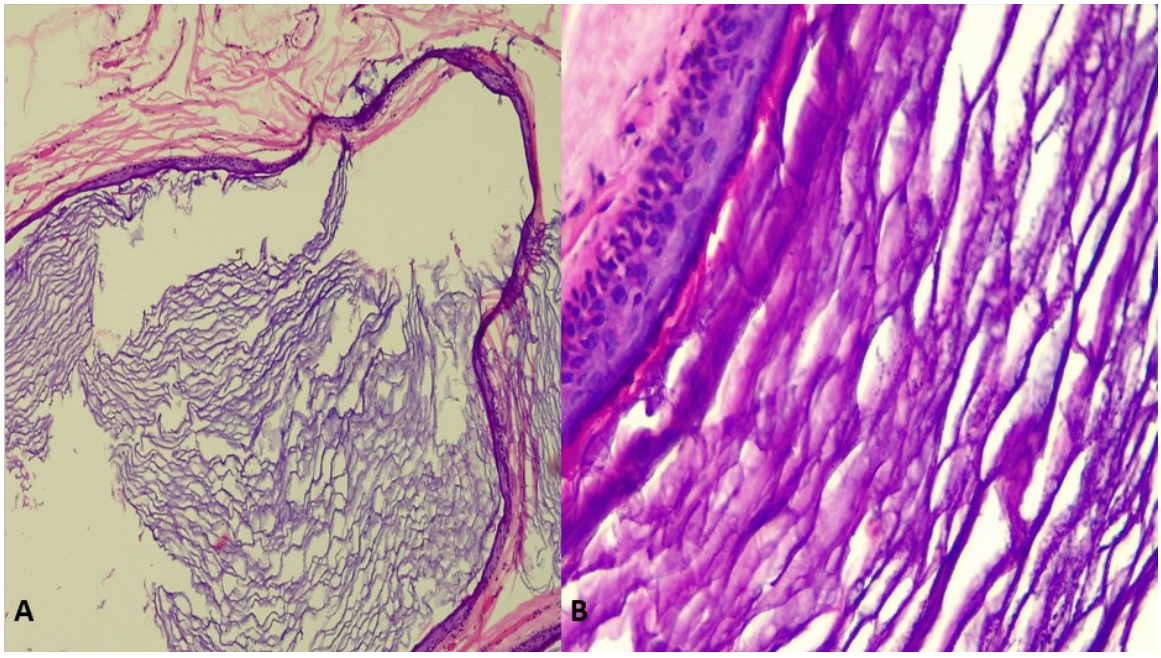

Figure 4 [A, B]: Photomicrographs of the submental mass; (**A**) shows a low power view of the cystic mass, displaying the thin cyst wall and the abundant keratin flake cystic content [H&E, X40 mag.], (**B**) shows a high-power view of the cyst wall and its adjacent keratin flakes [H&E, X400 mag.].

of lower jaw (submental) swelling, which was painless, insidious in onset, initially very small, but gradually increased in size over the years to attain its present huge size. There was no history of preceding trauma to the site, oral cavity or neck (including previous surgeries in the submental region). She also had no history suggestive of dryness of the mouth (xerostomia), dysphagia, change in voice, difficulty in breathing, or other swellings in the mouth or any other part of her body; with no family history of similar illness. She neither consumes alcohol nor tobacco (in any form). She has not received any form of treatment for this lesion; neither medications nor surgery, such as local incision and drainage (I&D). Patient lost her mother in her early childhood, while her father is still alive and works as an artisan (daily worker). She became seriously concerned with her disfigured facial appearance as a young lady, hence she decided to seek for surgical therapy (cosmesis) at this time.

On examination, she was found to be a young lady, that is 1.62 meters tall and weigh 56.8 kilograms; with a calculated body mass index (BMI) of 21.68kg/m2. She was not in any obvious painful or respiratory distress. There was a huge submental mass measuring 8.0 x 10.0cm, across its widest dimensions, see **Figure 1**. This submental mass neither moved with swallowing nor with the protrusion of the tongue. The skin over this submental mass appeared healthy with no scarification marks or engorged vessel. This mass was non-tender, firm in consistency, and non-adherent to underlying and overlying structures. On the inspection of the oral cavity/oropharynx, no mass lesion was conspicuous on the floor of the mouth, however on bimanual palpation of the floor of the mouth, a huge smooth walled firm mass was felt.

The ultrasound scan (USS) of the neck carried out, suggested a hypoechoic cystic lesion of the submental region with no evidence of malignancy. We also requested for other radiological investigations to further characterise this lesion, namely computed tomography (CT) scan and magnetic resonance imaging (MRI), but the patient could not pay for these tests because she was indigent and was not under any health insurance cover; she could barely afford the surgical procedure cost.

A working diagnosis of a giant plunging ranula was made, and patient was worked up for surgical excision (as a corrective surgery) of a plunging ranula the following week.

We requested for the routine blood investigations essential as pre-operative requirements for this corrective surgery, and the investigation results were within normal range.

We performed this corrective surgery under general anaesthesia through an oro-endotracheal intubation, and a transverse submandibular incision was made to divide the skin. To gain further entry into the submental region, subplatysmal muscle flaps were raised and the mylohyoid muscle was dissected to reveal the submental mass lesion. Intra-operatively, we found a huge ovoid well circumscribed cystic mass located just adjacent to her left submandibular salivary gland, while extending from the left mandible to the midline, see **Figure 2 [A – C]**. We ensured oral mucosal integrity by examining the floor of the mouth for any breach that may require a repair. We immediately sent this excised mass for histopathological evaluation.

We closed the surgical wound in layers, using absorbable sutures for the subcutaneous layer and non-absorbable sutures for the skin, and subsequently applied pressure dressing to the site. Furthermore, we placed her on parenteral antibiotics and non-steroidal anti-inflammatory drugs (NSAIDs) for her post-operative care. We removed her skin sutures/stitches after 5 days. The surgical wound site was found to be healthy and thus patient was discharged to go home, and given an ENT outpatient clinic visit appointment for follow up.

At surgical cut-up, the specimen was found to be an ovoid tan-grey fluctuant cystic mass with a smooth capsule; measuring 9.0 x 6.0 x 5.0cm in its widest dimensions and weighing about 20grams. Transection of the specimen revealed a unilocular cystic cavity containing about 20mls of tan-grey to cream coloured soft semi-solid material. The inner cyst lining was smooth with variable cyst wall thickness, ranging from 0.1 to 0.2cm, see **Figure 3**. Representative sections were taken from the specimen and submitted for tissue processing and microscopy. On microscopic evaluation, sections from this cystic mass wall showed a variably thin fibrocollagenous cyst wall lined by atrophied

stratified squamous epithelium displaying marked orthokeratosis, hyperkeratosis and a cystic cavity filled with keratin flakes. Skin appendage and adnexae were absent, and also other tissues that characterise multiple germline origin were absent, hence ruling out dermoid and teratoid cysts diagnosis, see **Figure 4**. We made a histopathologic diagnosis of submental epidermoid cyst (epithelial inclusion cyst) based on these gross and microscopic diagnostic features/criteria.

Discussion:

In 1955, Dr. Meyer in his study of cysts of the mouth, histologically classified them into three groups, namely: epidermoid, dermoid and teratoid.[13] By definition, epidermoid cysts are cysts whose inner surfaces are lined by stratified squamous epithelium devoid of skin adnexae/appendages, while dermoid cysts are cysts whose inner surfaces are lined by stratified squamous epithelium and additionally have skin adnexae/appendages, and teratoid cysts are cysts that have tissue components arising from ectoderm, mesoderm and endoderm such that in addition to the squamous epithelium and associated skin adnexae, tissues such as bone, cartilage, teeth, hair, fat, etc may also be present.[2,3,6,7,13] Though there are conflicting data on the commonest of these three types of cysts particularly between epidermoid and dermoid cysts, however most studies agree that epidermoid cysts comprise the commonest type.[2,4,7]

Epidermoid cysts can arise from variable aspects of the oral cavity, namely: sublingual, submental, submandibular, labial/lingual, buccal mucosa and tonsils.[2,3,14] The commonest site is the sublingual region (floor of the mouth) and the rarest site is the tonsillar region.[2,14] Furthermore, these epidermoid cysts can be categorised into three anatomical groups, namely: sublingual (medial genioglossus), submental (medial geniohyoid) and lateral (superior to mylohyoid muscle).[8,15,16]

Submental epidermoid cyst, as found in our index case, is indeed a very rare pathologic entity of the oral cavity particularly and of the head and neck region generally, given that it accounts for less than 0.01% of oral cavity cysts.[2–5,7,15–20] Similarly, in the head and neck region, the incidence of epidermoid (and dermoid) cyst ranges from 1.6% to 6.9% worldwide.[3,5,7,8,14] Additionally, submental epidermoid cysts occur most commonly amongst individuals aged 15 to 35 years, being very rare in children, and most common in males; our index case's age is within this commonest age range, though a female.[2–4,9,18,20]

The aetiopathogenesis of submental epidermoid cyst remains largely unknown, however there are three theories still under consideration, namely: dysontogenetic or dysembryogenetic (defective development of an embryo), traumatic and thyroglossal anomaly.[3,15,18] Furthermore, epidermoid cyst can be categorised, based on aetiology, into two groups, namely: congenital and acquired.[1,3–5,15] Congenital epidermoid cyst arise as dysembryogenetic lesions which are formed due to entrapment of ectodermal elements by first and second branchial arches during their midline fusion in the third to fourth weeks of intrauterine embryonal development. This congenital epidermoid cyst may also arise due to abnormal formation of the tuberculum impar of His, which gives rise to the floor of the mouth, the body of the tongue and the mandibular arch. Likewise, congenital epidermoid cyst may also present as a variant of thyroglossal duct cyst.[3] Conversely, acquired epidermoid cyst arise due to traumatic events (such as surgery), iatrogenic inclusion of epithelial cells during medical procedures, and occlusion of the duct of a sebaceous gland respectively.[3,5] The aetiology and aetiopathogenesis of the submental epidermoid cyst in our index case was most probably in accordance with the dysembryogenetic theory, and hence of the congenital category given that the lesion has been present for 10 years.

Epidermoid cysts of the oral cavity grow slowly and progressively as a midline structure in the floor of the mouth and may eventually displace the tongue presenting as a painless soft fluctuant mass lesion and causing varying degrees of difficulties in swallowing (dysphagia), speaking (dysphonia), breathing (dyspnoea) and may eventually give rise to a characteristic double chin appearance with lower localization.[2–4,7–11,15,16] The index case had this characteristic double chin appearance because of the submental localization of the mass lesion.

Radiological investigation modalities such as ultrasound scan (USS), computed tomography (CT) scan and magnetic resonance imaging (MRI), in

addition to histopathological evaluation, are integral to diagnostically characterising epidermoid cysts as well as making adequate surgical management plan for it.[21–25] USS of submental epidermoid cyst will characteristically show a well circumscribed midline cystic ovoid to spherical, lobulated or tubular predominantly hypoechoic to hyperechoic (usually multiple rounded echogenic) structure(s) with no significant vascularity with dorsal or posterior acoustic amplification and lateral shadowing with or without scattered internal dark clefts giving a pseudocyesis pattern appearance; which presents as anaechoic for very small lesions and heterogenous for very large lesions as well as displaying twinkling artefact on colour doppler USS.[21–25] CT scan of submental epidermoid cyst will characteristically show a well encapsulated mass of heterogenous densities (hypodense submental mass) that represent a mixture of fat and keratin which can calcify within; having a density similar to that of water with slender and sclerotic margins, presenting as a homogenous fluid attenuation non-enhancing mass lesion.[21–25] MRI of submental epidermoid cyst will characteristically show a mass with mild hypointense signal intensity on T1-weighted (T1W) and intermediate to high signal on T2-weighted (T2W), some true restricted diffusion and superimposed T2 shine through on DWI, and no enhancement centrally with or without thin peripheral enhancement on T1 C+ (Gd) images. This is so because this lesion on MRI have a density similar to water or cerebrospinal fluid (CSF) following fluid signal intensity.[21–25] It is of note that while contrast-enhanced CT scan is a faster and cheaper modality than contrast-enhanced MRI studies, MRI is better in delineating the soft tissue and vascular aspects of submental epidermoid cyst.[21] Though these radiological modalities discussed above are invaluable in the management of submental epidermoid cyst, our patient could only afford to pay for USS which showed a hypoechoic cystic lesion.

The differential diagnosis of oral (in this case, submental) epidermoid cyst includes: salivary retention cyst or ranula (including giant plunging ranula), unilateral or bilateral blockage of Wharton's ducts, lipoma, thyroglossal duct cyst, cystic hygroma, branchial cleft cyst, acute infection or cellulitis of the floor of the mouth, median neck cyst, lymphadenopathy, submental abscess, Sialolithiasis, Sialadenitis and/or infections of submandibular and sublingual salivary glands, odontogenic abscess, mucus extravasation, Haemangioma, floor of the mouth and adjacent salivary gland benign and malignant neoplasms, heterotopic gastrointestinal cyst and duplication foregut cyst.[2,3,15–18,20] Our index case was first thought to be a giant plunging ranula, however this diagnosis changed to submental epidermoid cyst following histopathological evaluation of the mass sent to the histopathology department of our hospital after a successful surgery. This kind of scenario of masquerading lesions has been well reported by similar case reports.[10–12,15,17,26] Histologically, a plunging ranula shows a psedocystic (devoid of epithelial lining) lesion lined by muciphages (epithelioid macrophages that have engulfed extravasated mucin) which is in contrast to an epidermoid cyst lined by stratified squamous epithelium.[1,12] Thus, buttressing the need to have high index of suspicion in dealing with such cases and also to emphasize that histopathological evaluation should serve as the gold standard for definitive tissue diagnosis in medicine and surgery.[8,10–12,14–19] Furthermore, given the possibility of these oral cystic lesions to transform into squamous cell carcinoma and basal cell carcinoma in its natural history, it is very important to adequately follow-up these patients whenever a tissue diagnosis of epidermoid cyst in surgical lesions of the floor of the mouth is made.[2,15–17] The good news, however, in this lesion is that the probability of this change (as well as recurrence of the cyst) is low and that surgical excision, which was performed in our index case is curative and effectively removes this possibility of malignant transformation.[2,15–17] Also, the surgical procedure in the index case was through the extra-oral route and this approach has been reported as the best way to operate on submental mass lesions in other similar studies.[15,17,18]

In conclusion, submental epidermoid (epidermal) cyst is a very rare non-neoplastic cystic lesion of the oral cavity which can mimic giant plunging ranula in its clinical presentation. Histopathologic evaluation is crucial in making definitive tissue diagnosis in such scenarios, and this will guide expert management of these patients. Further

research into the definitive aetiopathegenetic pathway in view of its malignant transformation potential is hereby recommended.

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