

Case Report

A case of an unusually large Sublingual dermoid cyst of the maxillofacial region

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Abstract

Dermoid cysts are true developmental cysts, which arise from the entrapment of pluripotent ectodermal and mesodermal primordia in the embryonic lines of fusion, between the 3rd and 5th weeks of gestation. Hence, they are dysontogenic in origin, that is, they result from defective embryonic development. In the head and neck region, these ectodermal and mesodermal components get entrapped when the 1st and 2nd branchial arches of each side fuse in the midline. The entrapped tissues then undergo proliferation and cystic transformation. A case of a large sublingual dermoid cyst in a 37 year old male patient is presented which closely mimicked a plunging ranula and was successfully managed by surgical excision.

Keywords: Dermoid, Epidermoid, Teratoid, Ranula.

INTRODUCTION

Dermoid cysts are “Choristomas” as well, i.e., tumors or pathologies originating from the proliferation of aberrant primordial tissue not native to that part. In other words, they are composed of normal appearing tissue in an abnormal location.

Dermoid cysts are relatively uncommon in the Oral and Maxillofacial region, accounting for just 2-7% of all dermoids and comprising about 34% of all developmental cysts of the head and neck. In this region they are mostly found in the periorbital lateral eyebrow area, the “orbital dermoid cysts”, followed by the submental region, the “submental dermoid”, external to the mylohyoid muscle or in the floor of the mouth, the “sublingual dermoid”, oral to the mylohyoid muscle. Others are the “nasal dorsum dermoid cysts” arising from inclusions between the developing nasal bones.

A case of a large sublingual dermoid cyst in a 37 year old male patient is presented which closely mimicked a plunging ranula and was successfully managed by surgical excision.

Case Report

A 37 year old male patient presented with a painless swelling below the chin over the front of the neck, which had been present for the past two years, slowly increasing in size. Since the past 6 months, it had caused some difficulty in speech. On examination, there was a single, localized, sessile, spherical swelling in the anterior midline of the neck, measuring 8x8cm and extending from the inferior border of the mandibular symphysis to the level of the cricoid cartilage (Figure 1). The surface of the swelling was smooth, borders indistinct and the overlying skin appeared normal. The swelling was cystic in consistency, bimanually palpable, compressible but non reducible, with no evidence of inflammation, fluid thrill or impulse on coughing. The tongue and floor of the mouth were raised, by a solitary, well circumscribed, distinct, dome shaped sessile midline swelling extending from the lingual aspect of the mucogingival junction of mandibular anterior teeth up to the mandibular molars bilaterally (Figure 2). The mucosa over the swelling appeared normal without any secondary changes. The morphology of the swelling did not vary with tongue movement. On palpation, the swelling was soft to firm,

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Figure 1a and b. A 8x8 cm swelling in the submental region of a 35 year old patient



Figure 2a and b. Close resemblance to a plunging ranula. The large sublingual swelling raised the tongue upwards. Also seen are the punctae of the Wharton's ducts.



Figure 3a,b and c. Axial, Sagittal and Coronal sections of the CT scan revealed a large well circumscribed radiolucency in the submental region displacing the tongue upwards and causing a narrowing of the upper airway space.

non tender, smooth, fluctuant and was not associated with any discharge. Submandibular and sublingual gland orifices appeared normal and bilateral milking of the glands produced thick, mucous saliva. The clinical impression was that of a sublingual dermoid cyst, while the differential diagnosis included a plunging ranula, epidermoid cyst, thyroglossal duct cyst, branchial cleft cyst, cystic hygroma, submental/sublingual space

infection or lymph node enlargement, teratoid cyst, sublingual salivary gland infection or tumor.

Mandibular occlusal / cross sectional radiographs did not disclose any calcifications. The buccal and lingual cortical plates were normal with no indication of expansion or decortication. CT scans (axial, coronal and sagittal sections) revealed a large radiolucent space occupying lesion in the sublingual space (Figure 3).



Figure 4. Aspiration of the cyst yielded a straw coloured fluid containing yellowish-white, cheesy, granular material.

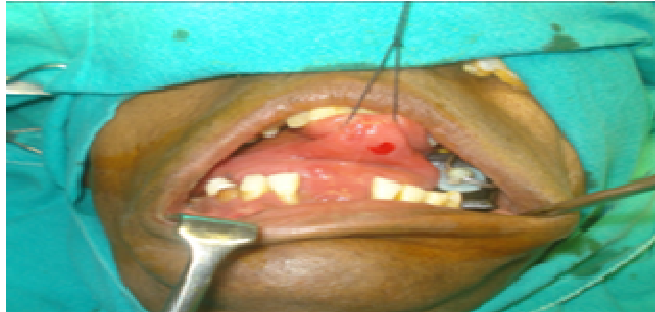


Figure 5. Upward retraction of the tongue with the aid of a silk suture.

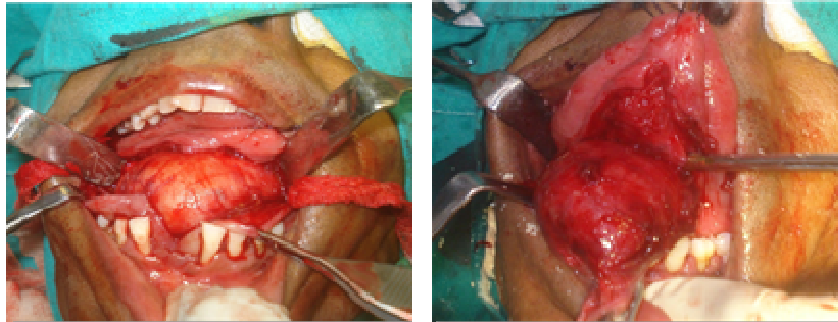


Figure 6 a and b. An intraoral, inverted T- shaped midline incision from base of the tongue to floor of the mouth, providing excellent exposure of the lesion, followed by blunt pericapsular dissection in an avascular plane.

Aspiration of the cyst yielded a semisolid cheesy material (Figure 4) , which on cytology, showed a few squamous cells visible in the background of sheets of macrophages, few lymphocytes, abundant keratin flakes and anucleate squames.

The patient was taken up for surgical enucleation of the cyst under GA. Adrenaline, 1:100,000 were infiltrated into the floor of the mouth. A suture was placed through the tip of the tongue, which was then retracted upwards

(Figure 5). An inverted “T” shaped incision was made in the midline across the anterior part of the swelling, taking care to avoid any damage to the Wharton’s duct and its opening on both sides (Figure 6). Careful submucosal dissection revealed the large cyst, which was then enucleated (Figure 7, 8). The openings of the Wharton’s ducts were checked for patency followed by closure of the surgical site (Figure 9).

The resected specimen showed presence of

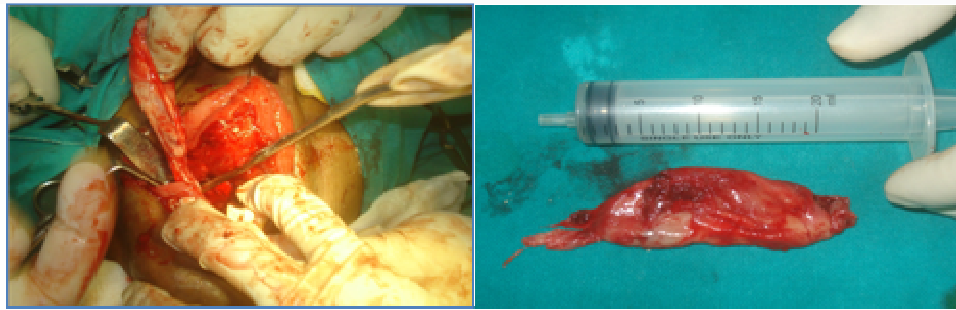


Figure 7. Total enucleation of the cystic lining.

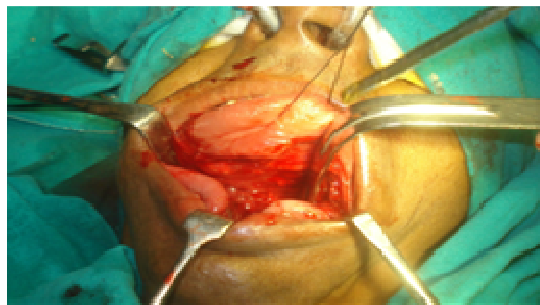


Figure 8. After cyst enucleation.

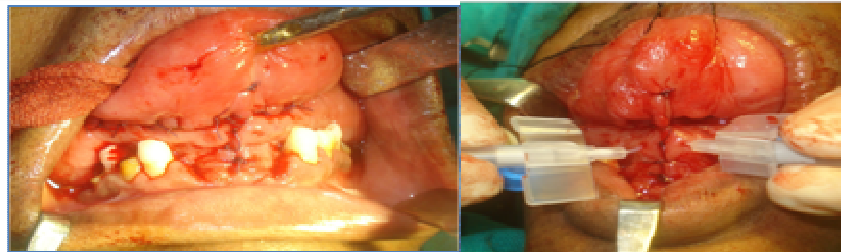


Figure 9. Checking for patency of the Wharton's ducts.

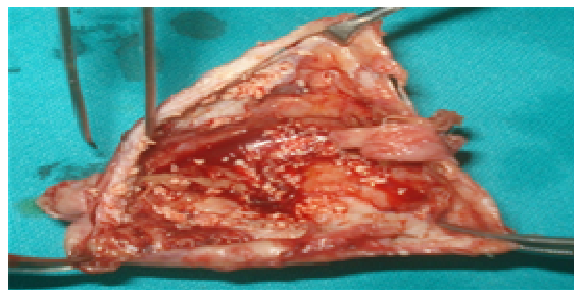


Figure 10. Cheesy keratinous material seen within the lumen of the excised dermoid cyst.

yellowish-white, cheesy, granular material within its lumen (Figure 10), which on histopathologic examination

was confirmed to be keratin flakes. Postoperative healing was excellent (Figure 11, 12). Histopathologic examina-



Figure 11. Good healing seen at the end of a week.



Figure 12. A complete resolution of the extraoral swelling after excision of the sublingual dermoid cyst.

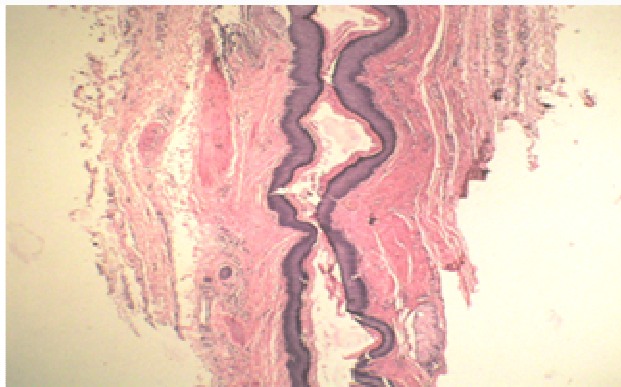


Figure 13. H&E Section 10X, showing keratinised stratified squamous epithelium lining the cyst lumen. Also seen, is a dense lymphocytic infiltration of the connective tissue wall.

nation of the excised specimen showed that the epithelial lining was of the keratinising, stratified squamous type and the connective tissue wall showed the presence of lymphocytes and multinucleated giant cells with deeper

tissue fibrosis and prominent blood vessels, and a number of sebaceous glands (Figure 13, 14). Hence, it was conclusively diagnosed as a dermoid cyst.

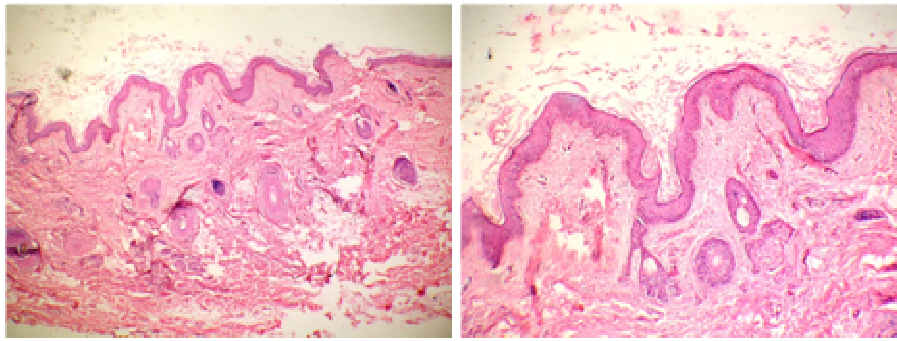


Figure 14. H&E Section 10X and 20X, showing shredded keratin flakes desquamating from the keratinized stratified squamous epithelial lining of the dermoid cyst. Also seen are proliferating sebaceous glands embedded within the fibrous connective tissue wall.

DISCUSSION

The word "Dermoid cyst", is a generic term which has been used to encompass and describe three different histological varieties. All three types may contain a cheesy keratinous material. Based on the histopathologic picture, Meyer divided the floor of the mouth cysts into following types: (Black et al., 1993; Janjua and Goravalingappa 1999; Teszler et al., 2007).

a) Epidermoid cysts, which are simple cysts, lined with simple squamous epithelium, with a fibrous wall and no skin appendages or adnexals like hair follicles, sebaceous or sweat glands in their connective tissue wall. As these cysts develop from the upper part of the pilosebaceous unit, they are incapable of producing sebum and do not contain any skin appendages.

b) True Dermoid cysts, also known as compound cysts, which are lined with keratinizing stratified squamous epithelium with skin adnexals in the connective tissue wall. The lumen will contain keratin, sebum, varying amounts of fat and occasionally, hair.

c) Teratoid cysts, also known as complex cysts, which are lined with epithelium ranging from simple stratified, keratinized stratified squamous to stratified columnar respiratory type of epithelium and containing derivatives from all the three germ layers (ectoderm, mesoderm and endoderm) within their lumen. The cystic cavity in addition to skin appendages also encloses mesodermal derivatives such as bone, muscle, gastrointestinal and respiratory tissue.

Theories of etiology of dermoid cysts:

- i) Congenital inclusion of dermal and epidermal elements of germ layers in deeper tissues along the embryonic lines of fusion.
- ii) Acquired traumatic implantation of dermal and epidermal elements of surface epithelium, which may proliferate and keratinize.
- iii) Growth from rest of totipotent cells displaced from the blastomere.

Although floor of the mouth in the midline is most favoured site, occasional occurrence involving the buccal mucosa, tongue, lips, uvula, temporomandibular joint dermal graft, intradiploic, intracranial, and intraosseous location within the mandible and maxilla also have been cited in literature (Koca et al., 2007; Hemaraju et al., 2004; Shear and Speight, 2007). These lesions show variation in size and weight from few millimetres to centimetres and a gram to several hundred grams respectively (Teszler et al., 2007; Pancholi et al., 2006).

Dermoid cysts present as a single, localized, slow growing, mobile, soft, firm or doughy, usually well circumscribed soft tissue mass. In the head and neck region, they are mostly seen in the midline and even when they are laterally positioned; their stalk can often be traced to the midline. They are usually seen in children, adolescents or young adults. They may present later in life as they are slow growing and become evident only after reaching an appreciable size. They may at times show a sudden increase in size during puberty or pregnancy, due to the hormonal changes (increased plasma levels of estrogen and progesterone), which stimulate the sebaceous glands that these cysts contain, with a resultant abundant sebum secretion and accumulation.

The sublingual dermoids raise the tongue and may cause difficulty in speech, swallowing and breathing, while the submental dermoids produce the characteristic "double chin" appearance. Symptoms of dysphagia, dyspnoea and dysphonia may occur due to upward displacement of tongue by these sublingual swellings (Koca et al., 2007; Pancholi et al., 2006). Further more growth in a inferior direction may give rise to appearance of characteristic "double chin" (Black et al., 1993; Koca et al., 2007; Hemaraju et al., 2004). These well encapsulated lesions typically feel "dough like" on palpation, although they may be fluctuant and cyst like based on consistency of the luminal contents, that may

range from a cheesy, sebaceous to liquefied substance (Pancholi et al., 2006; Kandogan et al., 2007).

On an ultrasound, they typically appear as a well circumscribed, thin walled, unilocular mass with coalescence of fat globules into small, discrete, echogenic nodules within the fluid matrix, producing a characteristic "sac-of-marbles" appearance. On histopathologic examination, dermoid and epidermoid cysts are lined with stratified squamous epithelium with keratin and / or sebaceous material filled cavity. In a true dermoid cyst, the connective tissue wall shows variable dermal appendages, including sebaceous glands, hair follicles and sweat glands.

The differential diagnosis for sublingual dermoids comprise ranula, unilateral or bilateral blockage of Wharton's ducts, lipoma, thyroglossal duct cyst, cystic hygroma, branchial cleft cysts, acute infection or cellulitis of the floor of the mouth, infections of submaxillary and sublingual salivary glands, floor of the mouth and adjacent salivary glands benign and malignant tumours, heterotopic gastrointestinal cyst and duplication foregut cyst (Black et al., 1993; Janjua and Goravalingappa 1999; Koca et al., 2007; Kandogan et al., 2007; Hemaraju et al., 2004).

Treatment comprises total surgical excision (Black et al., 1993; Janjua and Goravalingappa 1999; Koca et al., 2007; Kandogan et al., 2007). Caution should be taken not to rupture the cyst, as cystic contents may act as irritants to fibrovascular tissues, causing postoperative inflammation (Teszler et al., 2007). Recurrences are unusual after absolute surgical excision (Black et al., 1993; Teszler et al., 2007). Reports of malignant transformation of sublingual dermoid and epidermoid to squamous carcinoma and basal cell carcinoma are present (Rutherford et al., 2006; Mustafa et al., 2003). A 5% rate of malignant transformation of the teratoid variety of oral dermoid cysts has also been quoted in literature

(Shear and Speight, 2007; Rutherford et al., 2006; Mustafa et al., 2003).

CONCLUSION

Dermoid cysts of the maxillofacial region are relatively uncommon entities. Ample understanding and vigilance about these slow growing painless masses is essential not only because of the symptoms they produce but also due to their potential for recurrence.

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