Potentially Fatal Supramylohyoid Sublingual Epidermoid Cyst

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Abstract A case of chronic and slow growing massive lateral neck swelling is presented which gradually resulted in dysphagia to an extent that patient reported in emergency room. Clinical findings were indicative of a cystic swelling or a massive lipoma. Temporary decompression of the lesion was achieved by partially aspirating the contents of the cyst. Nature of aspirate and its microscopic and biochemical analysis excluded lipoma, vascular malformation and salivary phenomenon. The diagnosis tapered to developmental lateral neck cysts. Magnetic Resonance Imaging (MRI) revealed a massive cystic lesion in the left floor of mouth extending to the right lingual aspect of mandible and posteriorly to impinge on the medial wall of pharynx. A combined intraoral and extraoral approach was used to expose and excise the lesion in toto. Final histological diagnosis of the pathology was epidermoid cyst.

Keywords Dermoid cyst · Dysphagia · FNAC · Lateral neck swelling

Introduction

Epidermoid cysts are circumscribed; slow growing and asymptomatic cysts of soft tissues. They go unnoticed until they are visually apparent as swelling. This is the reason for patients to present in 2nd or 3rd decade. There is no gender predilection [1]. In case of superimposed infection, they might form a draining sinus or may cause fatal airway compromise [2]. Pain and other inflammatory signs may set in at this juncture.

Epidermoid cysts are a histological variant of dermoid cysts. Dermoids have three histological subtypes, namely; true dermoid cysts, epidermoid cysts and teratoid cysts. These cysts are lined by stratified squamous epithelium. True dermoids show skin adnexae in surrounding wall namely sebaceous and sweat glands and hair and hair follicles. If the cyst does not contain dermal appendages, the variety is called epidermoid cyst. Teratoid cyst contains elements derived from all three germ layers; ectoderm, mesoderm and endoderm, forming bone, muscle, gut or respiratory mucosa. The cysts are filled with muddy material composed of desquamated cells. Epidermoid cysts may sometimes show presence of secretory or ciliated epithelium which is why mucous has been found in some cases reported in literature [2, 3].

Case Report

A 16 year old male patient presented in casualty with a chief complaint of progressive difficulty in swallowing due to a swelling in left neck region. The resident on duty received a call for a patient with Ludwig angina. On examination, there were no signs of acute infection. Patient had noticed swelling about 3 months back when it was

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much smaller in size. The mass had gradually increased in size and was now causing difficulty in swallowing. There was no significant medical history and the general condition of the patient was normal except 2–3 episodes of intermittent low grade fever in past 3–4 months.

On examination, the swelling was 10×7 cm, extending from lower border of mandible to the level of inferior margin of thyroid cartilage (supero-inferiorly) and from anterior border of left sternocleidomastoid to contra lateral submental region (antero-posteriorly). Overlying skin was normal in colour, texture and temperature. The mass was not fixed to skin or underlying structures. It was compressible on palpation and mild tenderness could also be elicited. There was no history of discharge or associated sinus in relation to the swelling. Submandibular lymph nodes could not be palpated on ipsilateral side due to size of the lesion. Contra lateral submandibular nodes were enlarged but normal in other parameters. All other cervical nodes were normal. Neck movements were fine and cranial nerve functions were intact. Mouth opening was normal. Floor of mouth was raised on ipsilateral side. Intraoral hard and soft tissue examination was normal. There was no history of pain or difficulty in mastication or change in voice. Laryngeal crepitus was normal. There were no associated symptoms of dyspnoea, cough or throat pain. Mild dysphagia was reported since last 10 days. Patient negated history of any neck surgery or trauma. The swelling was immobile on tongue protrusion and swallowing (Fig. 1).

Aspiration resulted in dirty white thick fluid with dull whitish solid flakes that obstructed the wide bore needle lumen (18 G). Lesion could be decompressed only partially and temporarily on repeated attempts. Microscopic examination of the aspirate identified the material to be keratin. Culture was negative for bacterial growth. Biochemical

analysis was negative for salivary amylase. Magnetic Resonance Imaging of the neck revealed $6.2 \times 6 \times 5.7$ cm sized well defined, well margined cystic lesion in left sublingual space containing internal debris and few calcified areas. The mass had displaced the tongue supero-laterally and was encroaching on the pharyngeal wall medially. Posteriorly, the lesion extended onto the carotid space but did not invade it. There was no communication with the pharynx. Inferiorly the mass was bounded by the mylohyoid muscle (Fig. 2). Routine blood and urine investigations were in normal range. No abnormality was detected on chest radiograph.

Clinical examination and history of presenting complaint hinted towards a chronic, slow growing lesion, a plunging salivary pseudocyst, a low-flow vascular malformation, a deep seated massive lipoma or a congenital neck cyst. Nature of aspirate and its microscopic and biochemical analysis tapered the diagnosis to developmental cystic lesion. Further investigations were only to aid in surgical planning and could not have had an influence on treatment plan which was surgical excision of the lesion. The patient posted for the same under general anesthesia. Submandibular approach was used to address the lesion extraorally. Unlike conventional submandibular dissection plane, the lesion could be seen only after dividing the mylohyoid muscle. Additional approach from floor of mouth was helpful in dissecting the supero-medial aspect. After excision of the lesion, excess mylohyoid muscle was excised and an attempt was made to close the muscle in anticipated normal anatomic position. A vacuum drain was placed followed by layered closure (Fig. 3). Histopathology revealed a thin flattened hyperorthokeratinized stratified squamous epithelium with a prominent stratum granulosum and cuboidal to columnar basal cells. An inflammatory response comprising of lymphocytes, plasma cells, histiocytes and numerous foreign body type giant cells was seen associated with loss of epithelium at foci. The cyst wall was thin and fibrocollagenous with numerous dilated blood vessels and few fibroblasts. Some thick walled arteries and nerve tissue were also observed. Cystic lumen showed numerous keratin flakes. A final diagnosis of epidermoid cyst was made (Fig. 4).

Discussion

Dermoid cysts are chronic, slow growing and non tender soft tissue cysts. Two main theories of origin of these lesions have been proposed. Developmental theory proposes entrapment of pluripotent cells during 3rd to 4th week of intrauterine life when fusion of contra lateral first or second branchial arches occurs. The other school of thought says that the cysts may arise from traumatic

Fig. 1 **a** Preoperative frontal view showing lateral neck swelling. **b** Preoperative intraoral view showing raised floor of mouth displacing the tongue

Fig. 2 **a** T1- weighted axial MRI scan showing slightly hypointense signal from a large circumscribed mass in the floor of the mouth. **b** T2- weighted axial MRI scan showing hyperintense signal from a large well defined mass in the floor of the mouth. **c** T2-weighted coronal MRI scan showing hyperintense signal from a large well defined mass in the floor of the mouth causing upward displacement of the tongue. **d** T2-weighted sagittal MRI scan showing hyperintense signal from a large well defined mass in the floor of the mouth

inclusion of ectodermal cells in subcutaneous tissues. The mode of trauma may be iatrogenic or accidental. Patients usually present late as the lesion causes aesthetic or functional problem for e.g. cutaneous swelling, draining sinus, swelling in floor of mouth or other intraoral site, speech and/or masticatory problem or challenged airway [1, 3].

These cysts are rare in head and neck region. They comprise less than 0.01 % of cysts of oral cavity. In a review of 1,459 cases of dermoid cysts, 7 % were found in head and neck region and only 1.6 % in oral cavity. Another review of 541 dermoids of head and neck region reported the percentage of intraoral dermoids to be 6.5 %. Amongst intraoral sites, floor of mouth is most common [4, 5]. Dermoid cysts mostly arise in embryonic fusion lines so they commonly present as midline swelling. However, their site of origin may not always be in midline. If they start growing laterally and breach the mylohyoid curtain partially or completely, they may present as lateral neck swelling. Occasionally, there might not be any breach, rather stretching/expansion of mylohyoid by the pathology, which may present as lateral neck swelling as in our case [1, 6, 7].

Epidermoid cysts of the neck have been classified into three categories [8]:

- A. Supramylohyoid (intraoral)
 - a. Suprageniohyoid

- b. Infrageniohyoid
- c. Sublingual

B. Inframyohyoid (cervical)

- a. Submental
- b. Submandibular

C. Peri and trans mylohyoid (intraoral and cervical)

- a. Submental transmylohyoid
- b. Lateral perimylohyoid and transmylohyoid

Supramylohyoid sublingual epidermoids originate above the mylohyoid, deep to submandibular duct and lingual nerve and anterior to the stylohyoid ligament. In our case, the cyst attained a large dimension without breaching the mylohyoid muscle. It acted as a tissue expander to increase the dimension of the adjoining muscle. The patient did not experience any problem at rest except unaesthetic swelling of neck. Being from a rural area and low socio-economic strata, the patient neglected it until functional impairment crept in. During swallowing, the volume of oral cavity decreases as the hyoid along with its attached musculature is pulled up. The cyst was bounded inferiorly by mylohyoid and the act of swallowing would have caused the fluid in anterior portion of the cyst to shift posteriorly. The posterior limit of the cyst was encroaching on the medial

Fig. 3 **a** Excised specimen. **b** Intraoperative extraoral view showing exposure of lesion. **c** Intraoperative intraoral view showing lesion and left sublingual gland

wall of the pharynx. Fluid shift during swallowing would have further inflated the posterior cyst, which compressed on the pharyngeal musculature causing dysphagia. This prompted the patient to seek medical attention.

Emergency presentation of the patient with non-inflammatory, large, chronic swelling which was more or less fluctuant suggested a developmental etiology or mucous extravasation or a gigantic lipoma. Immediate aspiration was done which served two purposes. Firstly, it could further narrow the diagnostic possibilities. Secondly,

when the aspirate was found to be fluid, aspiration helped in decreasing the volume of cystic cavity which relieved dysphagia. Microscopic examination of the aspirate revealed keratin. Thus the provisional diagnosis of dermoid cyst or a branchial cleft cyst was formulated. However, while aspirating, extreme caution should be exercised for sterilization and asepsis. Secondary infection may have adverse consequences. Literature supports this statement and also suggests that aspiration should only be done in hospital setting [9].

Fig. 4 Photomicrograph showing thin flattened hyperorthokeratinized stratified squamous epithelium and a thin collagenous connective tissue wall (H and E, $\times 100$); *Inset* higher magnification (H and E, $\times 400$)

MRI is a gold standard in planning surgical excision. MRI usually reports a well defined cystic lesion with iso or slightly increased signal intensity on T1-weighted images and significantly high signal intensity on T2-weighted images. Internal debris and calcifications in walls may also be apparent [10]. STIR sequences are helpful to differentiate the lesion from lipomas. MRI was done to localize the lesion in relation to adjacent structures. The site of the lesion hinted more towards a dermoid cause.

Once a provisional diagnosis of a dermoid is formulated, incisional biopsy is not recommended. There may be chances of secondary infection, collapse of cyst due to drainage of contents making the surgical excision difficult and formation of draining sinus in relation to biopsy site. As surgical excision is the treatment of choice, and recurrence is extremely rare, thus we followed same principle in management of the patient. The cysts can be removed with or without overlying skin. If the cyst has formed a draining sinus, the sinus tract along with surrounding entropic skin is included in excision. Inclusion of skin can be in punch or elliptical fashion. Intraoral small supramylohyoid cysts can be approached from floor of mouth but large submylohyoid and trans/peri mylohyoid cysts may require extraoral or combination of intra and extra oral approaches [7]. Considering the size of cyst, we opted for a combined extraoral-intraoral approach to gain adequate access. Absence of superimposed infection or draining sinus made sure that the plane of dissection was discrete. After dividing the platysma, anterior belly of digastric was seen overlying the mylohyoid muscle which

had expanded and also attained significant thickness probably due to chronic pressure of the lesion. Digastric was retracted anteriorly and mylohyoid incised to gain access to the pathology. The cyst was massive and had some adhesions in superior and medial region. This was addressed intraorally. After exposure, the cyst was delivered extraorally.

Conclusion

Epidermoid cysts are rarely seen intraorally. When present, they might go unnoticed and assume large dimensions to impinge on upper aerodigestive tract. Aspiration in hospital setting and contrast enhanced MRI are useful tools in diagnosis. Excision is the treatment of choice. Intraoral approach is the first preference, but massive cysts may require extraoral approach or combination of both approaches. Recurrence is generally not seen.

References

1. Marx R, Stern D (2003) Oral and maxillofacial pathology. Quintessence, Chicago
2. Cortezzi W, de Albuquerque EB (1994) Secondarily infected epidermoid cyst in the floor of the mouth causing a life-threatening situation: report of a case. *J Oral Maxillofac Surg* 52:762–764
3. Leveque H, Saraceno CA, Tang CK, Blanchard CL (1979) Dermoid cysts of the floor of mouth and lateral neck. *Laryngoscope* 89:296–305
4. New GB, Erich JB (1937) Dermoid cysts of the head and neck. *Surg Gynecol Obstet* 65:48–55
5. Taylor BW, Erich JB, Dockerty MB (1966) Dermoids of the head and neck. *Minn Med* 49:1535–1540
6. Al-khateeb TH, Al-Masri NM, Al-Zoubi F (2009) Cutaneous cysts of the head and neck. *J Oral Maxillofac Surg* 67:52–57
7. Longo F, Maremonti P, Mangone GM, De Maria G, Califano L (2003) Midline (Dermoid) cysts of the floor of the mouth: report of 16 cases and review of surgical techniques. *Plast Reconstr Surg* 112:1560–1565
8. Teszler CB, El-Naaj IA, Emodi O, Luntz M, Peled M (2007) Dermoid cysts of the lateral floor of mouth: a comprehensive anatomic-surgical classification of cysts of the oral floor. *J Oral Maxillofac Surg* 65:327–332
9. Carner H, Lampe H, Downing P (1996) Dermoid cysts of the floor of the mouth diagnosed by fine needle aspiration cytology: a case report. *Acta Cytol* 40:319–326
10. Suzuki C, Maeda M, Matsumine A, Matsubara T, Taki W, Maier SE, Takeda K (2007) Apparent diffusion coefficient of subcutaneous epidermal cysts in the head and neck comparison with intracranial epidermoid cysts. *Acad Radiol* 14:1020–1028