

Lingual dermoid cyst with congenital discharging sinus: a rare entity

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Abstract

Objective: There have been few reports of lingual dermoid cysts with a congenital discharging fistula. We report such a case, which mimicked lipoma on computed tomography, and we discuss relevant clinical and radiological features.

Method: We present a case report and a review of the English language literature concerning lingual dermoid cysts either mimicking lipoma or with a congenital discharging sinus.

Results: A 35-year-old man presented with a lingual mass associated with a congenital fistula in the midline of the tongue dorsum, which discharged abnormal secretions. Computed tomography images showed that the lesion was well defined and contained homogeneous fatty tissue giving computed tomography attenuation values of -120 Hounsfield units.

Conclusion: Lingual dermoid cysts may develop with a midline discharging fistula, and may exhibit radiological similarity to lipomas on computed tomography.

Key words: Tongue; Dermoid Cyst; Pathology; Tomography, X-Ray Computed

Introduction

Dermoid cysts are lesions consisting of displaced ectodermal structures positioned along lines of embryonic fusion. The cyst wall is characterised by epithelium-lined connective tissue, including skin appendages. The head and neck region is frequently involved, but intralingual dermoid cysts are extremely rare.^{1–3}

Clinically, the lesion may grow slowly with an intact capsule, or may rupture with discharge of its contents. Keratin, sebum and hair may be present inside the cyst, and various radiological manifestations have been observed. The lesion may present as a solid mass or may contain liquid and/or fatty tissue.⁴

This case report discusses the challenges and clues leading to diagnosis, and gives a short review of the literature. The presented case demonstrates that, although rare, lingual dermoid cysts may present with a congenital discharging sinus and may mimic a lipoma radiologically.

Case report

History and clinical presentation

A 35-year-old man presented complaining of recurrent swelling of the floor of the mouth, with intermittent abnormal secretion from a congenital dorsum sinus. The symptoms had become more prominent in the past eight years, prompting the patient to seek treatment. The patient reported no significant pain, numbness or functional disturbance.

Physical examination revealed a mucosal orifice located in the midline of the central third of the tongue dorsum

(Figure 1), which was found to be approximately 5 mm in depth on probing. No prominent excretion was elicited with pressure. The mouth floor was swollen, with the mucosa soft and non-tender on palpation. Routine blood tests revealed no remarkable findings.

Imaging

Computed tomography (CT) images showed an oval, encapsulated, well demarcated mass approximately $6 \times 5 \times 4$ cm in

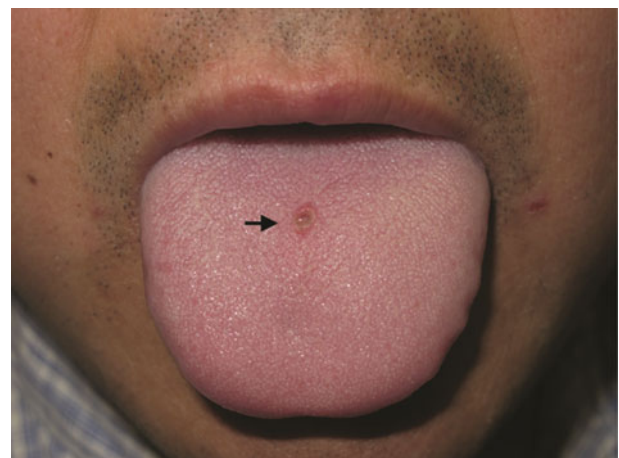


FIG. 1

Clinical photograph of the lingual dorsum sinus. Note that the opening of the sinus (arrow) is located in the central middle third of the midline, far from the foramen caecum.

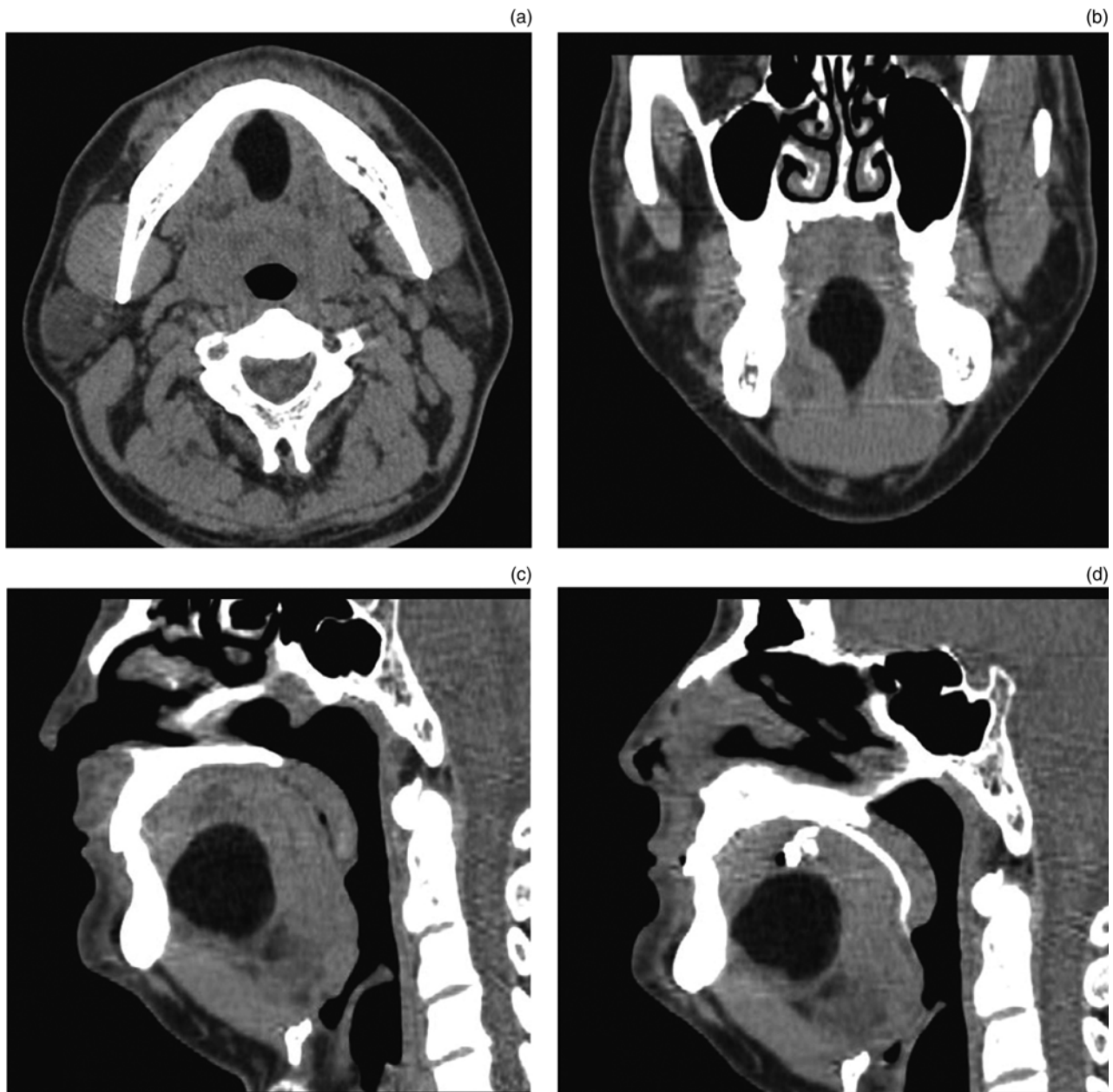


FIG. 2

(a) Axial, (b) coronal and (c) sagittal computed tomography (CT) images showing a well circumscribed lesion located within the genioglossus muscle and above the geniohyoid muscles, with a low attenuation value of approximately -120 Hounsfield units. (d) Sagittal CT sinograph showing a tract extending to the superior border of the cyst wall.

size, located in the midline of the tongue within the genioglossus muscle and above the geniohyoid muscles (Figure 2a to 2c). The mass was homogeneous and the attenuation was approximately -120 Hounsfield units.

Computed tomography sinography was performed immediately after contrast media (Iopamiro, 370 mg 1/ml; Bracco, Milan, Italy) had been injected through the dorsum orifice. The sinograph showed that the sinus tract extended to the superior margin of the capsule (Figure 2d).

Ultrasonography showed that the thyroid gland was present in the normal position.

Surgical management

As this lesion was clinically diagnosed as benign in nature, an intraoral excision was performed. En bloc resection of

the mass and the sinus tract was performed under general anaesthesia. The mass was well circumscribed and easily dissected from the adjacent soft tissue. The mass was encapsulated by a dense, fibrous capsule. Yellowish, mucous sebum was present in the mass.

The course of recovery was uneventful.

Histopathological examination

The specimen was fixed in 10 per cent buffered formalin and embedded in paraffin for routine haematoxylin and eosin staining. Microscopically, the cyst wall was seen to be composed of keratinising, stratified squamous epithelium, with keratin present in the cyst cavity. Abundant sebaceous glands were present in the cyst wall. None of the major components showed any pleomorphism or immaturity (Figure 3).

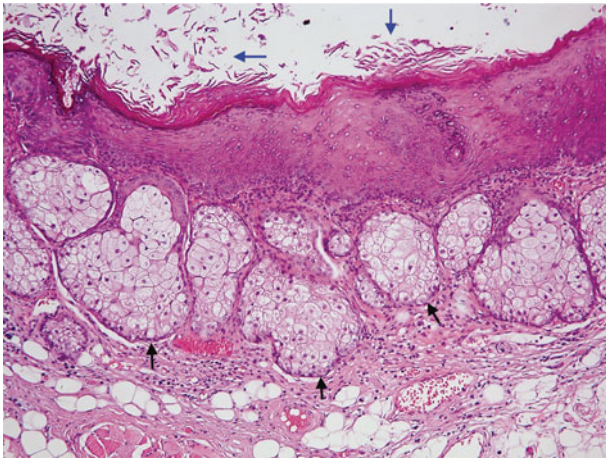


FIG. 3

Photomicrograph of the dermoid cyst showing the squamous epithelial cyst wall, which includes abundant sebaceous glands (black arrows); keratin is also seen within the cyst cavity (blue arrows). (H&E; $\times 200$)

The final diagnosis of dermoid cyst with congenital dorsum discharging sinus was confirmed.

Discussion

Embryologically, the tongue develops mainly from a pair of lateral lingual processes which project from the inner surface of the mandibular arch and fuse in the midline. The fusion site of the processes is the median sulcus of the tongue.

The pathogenesis of lingual dermoid cyst and dorsum sinus is closely related to embryogenic disorders of the tongue. Theories of pathogenesis include: epithelial entrapment and proliferation during fusion;⁵ failure of an embryonic cleft to disappear;⁶ differentiation of the displaced normal epithelial band into a serous gland; and traumatic implantation of epithelium.³ The congenital sinus in the midline of the dorsum may be derived from abnormal persistence of the epithelial band,⁷ or from a cyst rupture dissecting the tongue muscle along the line of least resistance.⁸ Confusingly, some authors have categorised dermoid cyst as a tumour. The presented case adds to our knowledge by suggesting that dermoid cysts may originate from abnormal embryogenesis rather than from an overgrowth of neoplastic cells. Thus, we believe that dermoid cysts should not be classified as tumours.

Thyroglossal duct cyst or sinus should be included in the differential diagnosis of a congenital tongue lesion with a sinus. Thyroglossal duct cysts and sinuses are caused by abnormal persistence or incomplete closure of the primitive thyroglossal duct. A thyroglossal duct cyst may occasionally develop within the tongue or in the floor of the mouth, and sometimes presents with a sinus extending to the foramen caecum. In our case, the sinus was located on the midline of the central third of the tongue, far from the foramen caecum. Thus, thyroglossal duct sinus could be excluded clinically.

The clinical course of a lingual dermoid cyst may be lengthy and the lesion may be symptomatic, depending upon its size. Difficulties in swallowing and sleeping may occur due to enlargement of the tongue.¹ Radiologically, dermoid cysts may appear as cystic, well demarcated lesions with fatty, fluid or mixed contents.⁹ Fatty tissue

components, where present, are specifically characterised by their low attenuation on CT images.¹⁰ In the present case, a CT attenuation value of -120 Hounsfield units strongly indicated the presence of a fatty component within the mass; this proved to be due to accumulated sebum from the abundant sebaceous glands within the cyst wall.

It is well known that lipomas typically exhibit low CT attenuation values of approximately -100 Hounsfield units. Lipomas can occasionally affect the tongue,^{11,12} and may be solitary, intermuscular or intramuscular.¹³ Although lipoma should be considered based on radiological findings, there have been no reports of recurrent swelling and abnormal discharge from a congenital sinus, due to a lipoma. Thus, it should be borne in mind that these two separate pathological entities may have great radiological similarity.

- **Lingual dermoid cyst is rare**
- **A congenital fistula may also be present discharging accumulated sebum**
- **In the presented case, computed tomography showed low-attenuation, homogeneous fatty tissue (resembling lipoma) due to sebum accumulation**

Secretions from the sebaceous glands accumulate primarily within the cyst, and are discharged when the cyst becomes too large to withstand compression from the tongue muscles. Rupture of a dermoid cyst may occur when continued expansion is anatomically restricted.^{14,15} In the presented case, the lesion developed in the midline of the tongue, within the genioglossus muscle. Muscle constriction during tongue movement made the lesion much more prone to rupture.

Dermoid cysts are sometimes associated with discharging sinuses, and can occur in the lip,¹⁶ nose,^{17–19} tongue,^{7,8} orbit²⁰ or other midline sites of the head and neck.^{21–24} In the present case, the external opening of the sinus was situated in the midline of the dorsum of the middle third of the tongue, and the sinus itself extended to the superior margin of the cyst wall. A CT sinograph showed that the sinus did not communicate with the cyst directly, indicating that occult rupture of the cyst wall may have been responsible for the observed discharge.

Surgical excision is the preferred management for lingual dermoid cyst.²⁵ Pre-operative evaluation of the location of the cyst in relation to the mylohyoid and geniohyoid muscles is important when deciding whether an intraoral or extraoral approach should be used for excision.⁵ The prognosis is usually good, and recurrence is rare after complete surgical excision.

Conclusion

Lingual dermoid cyst may present with a congenital, discharging sinus in the midline of the dorsum of the tongue, and may mimic lipoma on CT.

References

- 1 Dimtsas S, Theologie-Lygidakis N, Iatrou I. Intralingual dermoid cyst in an infant presenting swallowing and sleeping difficulties. *J Clin Pediatr Dent* 2010;**34**:335–7
- 2 Shaari CM, Ho BT, Shah K, Biller HF. Lingual dermoid cyst. *Otolaryngol Head Neck Surg* 1995;**112**:476–8
- 3 Miles LP, Naidoo LC, Reddy J. Congenital dermoid cyst of the tongue. *J Laryngol Otol* 1997;**111**:1179–82
- 4 Islam S, Hoffman GR. Parotid dermoid cyst: a rare entity. *J Laryngol Otol* 2009;**123**:e7

- 5 Meyer I. Dermoid cysts (dermoids) of the floor of the mouth. *Oral Surg Oral Med Oral Pathol* 1955;**8**:1149–64
- 6 Raewyn C, Paul W. Management of congenital lingual dermoid cysts. *Int J Pediatr Otorhinolaryngol* 2010;**74**:567–71
- 7 Giunta JL, Friedman AL, Karp R. Dermoid cyst of the tongue with sinus tract. *Oral Surg Oral Med Oral Pathol* 1982;**53**:450–3
- 8 Korchin L. Dermoid cyst with lingual sinus tract. Report of a case. *Oral Surg Oral Med Oral Pathol* 1974;**37**:175–8
- 9 Becker M. Oral cavity and oropharynx. In: Mafee MF, Vallavassori GE, Becker M, eds. *Imaging of the Head and Neck*, 2nd edn. Stuttgart, New York: Thieme, 2005;720
- 10 Mrad Dali-Grissa K, Zrig A, Mhiri-Souii M, Arifa-Achour N, Khochtali H, Tlili-Graies K. Intralingual dermoid cyst: imaging features of a giant cyst [in French]. *J Radiol* 2005;**86**:502–5
- 11 Dattilo DJ, Ige JT, Nwana EJ. Intraoral lipoma of the tongue and submandibular space: report of a case. *J Oral Maxillofac Surg* 1996;**54**:915–17
- 12 Chung JC, Ng RW. A huge tongue lipoma. *Otolaryngol Head Neck Surg* 2007;**137**:830–1
- 13 Nielsen GP, Mandahl N. *Lipoma*. Lyon: IARC, 2002
- 14 Smith AS, Benson JE, Blaser SI, Mizushima A, Tarr RW, Bellon EM. Diagnosis of ruptured intracranial dermoid cyst: value MR over CT. *AJNR Am J Neuroradiol* 1991;**12**:175–80
- 15 Kang MG, Kim KJ, Seok JI, Lee DK. Intracranial dermoid cyst rupture with midbrain and thalamic infarction. *Neurology* 2009;**72**:769
- 16 Kormanik KE, Bersu ET, Bentz ML. Dermoid sinus and cyst of the lip. *J Craniofac Surg* 2006;**17**:162–6
- 17 Cambiaghi S, Micheli S, Talamonti G, Maffei L. Nasal dermoid sinus cyst. *Pediatr Dermatol* 2007;**24**:646–50
- 18 Baarsma EA. The median nasal sinus and dermoid cyst. *Arch Otorhinolaryngol* 1980;**226**:107–13
- 19 Morimoto K, Takemoto O, Nishikawa M, Umegaki M, Nishino A. Nasal dermal sinus with a dermoid cyst. *Pediatr Neurosurg* 2002;**36**:218–19
- 20 Tien AM, Tien DR. Orbital dermoid cyst presenting with a discharging sinus tract. *J Pediatr Ophthalmol Strabismus* 2009:e1–2
- 21 Shen WC, Chiou TL, Lin TY. Dermal sinus with dermoid cyst in the upper cervical spine: case note. *Neuroradiology* 2000;**42**:51–3
- 22 Lee JK, Kim JH, Kim JS, Kim TS, Jung S, Kim SH *et al.* Cervical dermal sinus associated with dermoid cyst. *Childs Nerv Syst* 2001;**17**:491–3
- 23 Akhaddar A, Jiddane M, Chakir N, El Hassani R, Moustarchid B, Bellakhdar F. Cerebellar abscesses secondary to occipital dermoid cyst with dermal sinus: case report. *Surg Neurol* 2002;**58**:266–70
- 24 Layadi F, Louhab N, Lmejjati M, Aniba K, Ait Elqadi A, Ait Benali S. Cerebellar dermoid cyst with occipital dermal sinus. Report of two pediatric cases. *Pediatr Neurosurg* 2006;**42**:387–90
- 25 King RC, Smith BR, Burk JL. Dermoid cyst in the floor of the mouth. Review of the literature and case reports. *Oral Surg Oral Med Oral Pathol* 1994;**78**:567–76

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