J Oral Med Oral Surg 2025;31:35
https://doi.org/10.1051/mbcb/2025038
https://www.jomos.org
JOMOS
Journal of Oral Medicine and Oral Surgery

Case Report

Keratocystoma of palatal minor salivary glands: a case report

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(Received: 5 May 2025, accepted: 29 September 2025)

Keywords: Keratocystoma / choristoma / salivary gland tumor

Abstract – Keratocystoma is a rare benign tumor of the salivary glands. Initially described as a choristoma by Seifert *et al.* in 1999, the term "keratocystoma" was later proposed by Nagao *et al.* in 2001. The parotid gland is the most common site of occurrence. Only one case involving the minor salivary glands of the palate has been reported, by Ahuja in 2016. Herein, we present a rare case of keratocystoma arising in the palatal minor salivary glands of a 35-year-old man. Clinically, the lesion was painless, well-defined, with normal-appearing overlying mucosa, and located along the midline of the palate. These clinical features suggest a benign tumour of the minor salivary glands. Histological examination confirmed the diagnosis, revealing multiple cystic structures lined by non-keratinized squamous epithelium.

Introduction

Keratocystoma is an extremely rare, benign salivary gland neoplasm, first reported as a choristoma by Seifert *et al.* in 1999 [1]. The term "keratocystoma" was later coined by Nagao *et al.* in 2001 [2]. Most cases have been reported in the parotid gland [3]. Only one previous case affecting the minor salivary glands of the palate has been documented, by Ahuja *et al.* in 2016 [4,5]. This case report aims to present a second known occurrence of keratocystoma in the palatal minor salivary glands, highlighting its clinical, radiological, and histopathological characteristics.

Case report

A 35-year-old male presented with a painless, slow-growing palatal swelling, progressively increasing in size over the course of one year. He had no notable medical or surgical history. Intraoral examination revealed significant dental caries and residual root fragments.

Clinical evaluation showed a firm, painless, well-defined midline palatal mass measuring approximately 2 × 1.5 cm, with a sessile base and normal overlying mucosa (Fig. 1). Magnetic resonance imaging (MRI) revealed a solid lesion involving both the hard and soft palate, with mild post-contrast enhancement (Fig. 2).

The patient underwent surgical excision of the lesion under local anesthesia. A circumferential incision allowed for complete resection of the mass. A full-thickness mucosal flap was elevated to examine the underlying bone, which showed scalloping of the tumor bed. Curettage and superficial osseous resection were performed. Macroscopically, the excised nodule was well-circumscribed, homogenous, and whitish, measuring 2 cm at its greatest diameter.

Histopathological analysis revealed a nodular lesion composed of multiple cystic spaces lined by non-keratinized squamous epithelium overlying benign salivary gland parenchyma. The cysts contained lamellar keratin and scattered calcifications. The epithelium was regular, with both orthokeratotic and parakeratotic hyperkeratosis, and notably lacked a granular layer. Multinucleated foreign-body type giant cells of Müller type were observed surrounding keratin lamellae (Fig. 3).

These findings confirmed the diagnosis of keratocystoma of the palatal minor salivary glands. The postoperative course was uneventful, with satisfactory healing observed at the 10-day follow-up.

Discussion

Keratocystoma is a newly characterized, rare benign salivary gland neoplasm. Fewer than 15 cases have been reported in the literature to date [3]. All previously confirmed cases have involved the parotid gland, with a single exception reported by Ahuja *et al.* in 2016, involving the palatal minor salivary glands in a 34-year-old woman [4].

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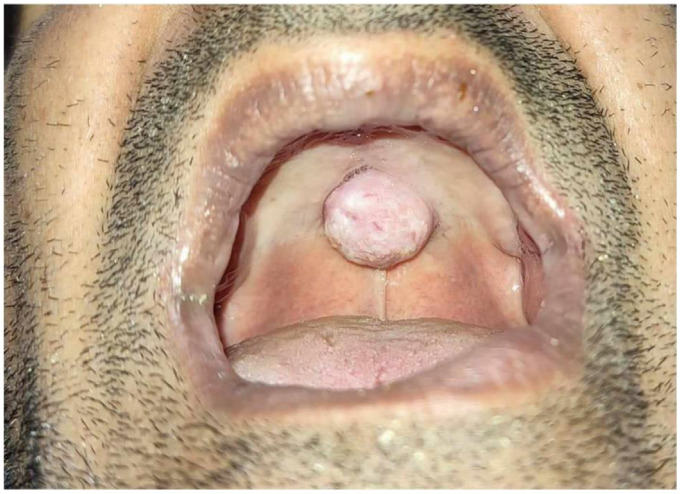

Fig. 1. Intraoral appearance of the palatal lesion.

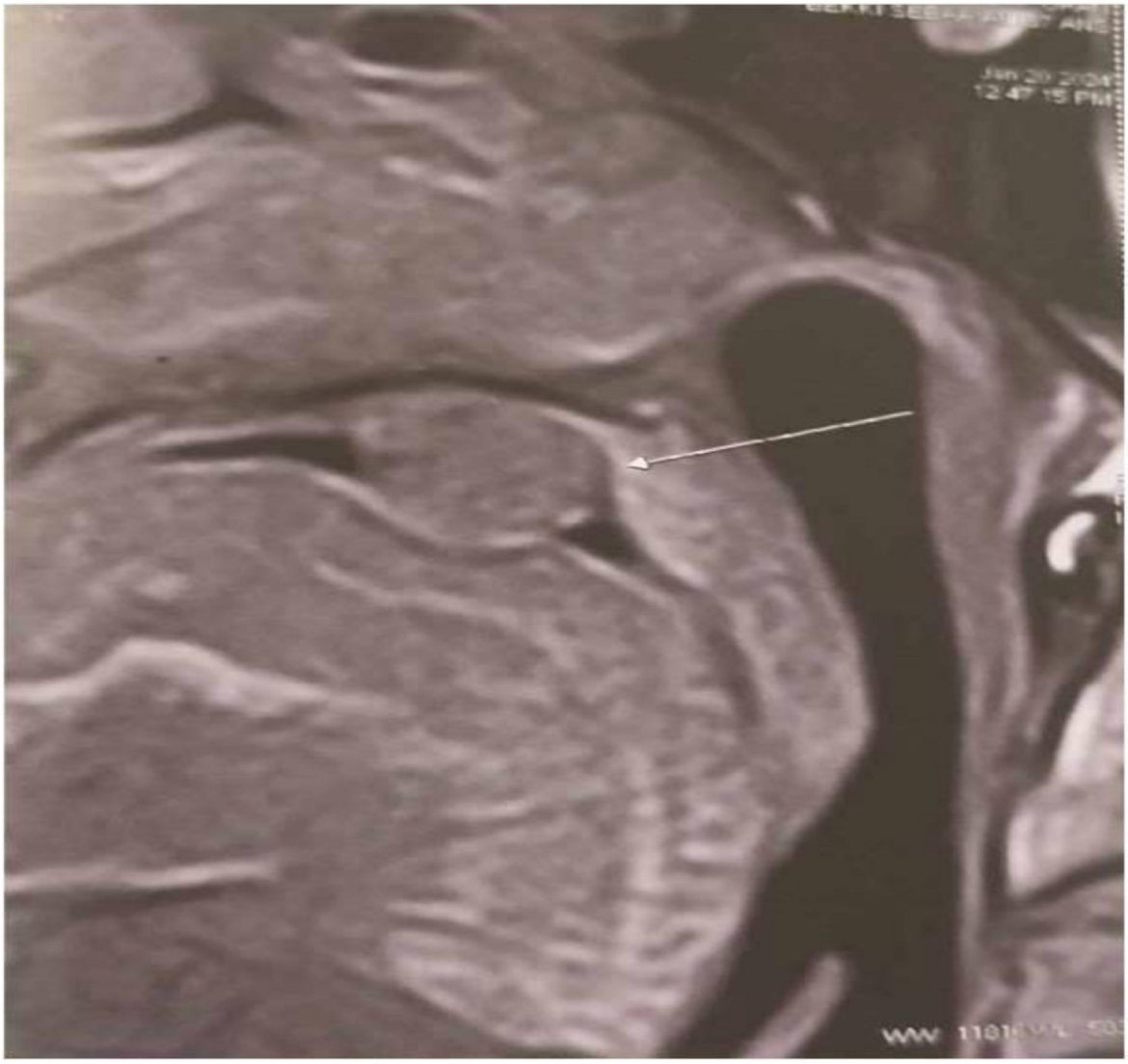

Fig. 2. MRI demonstrating contrast uptake and osseous scalloping.

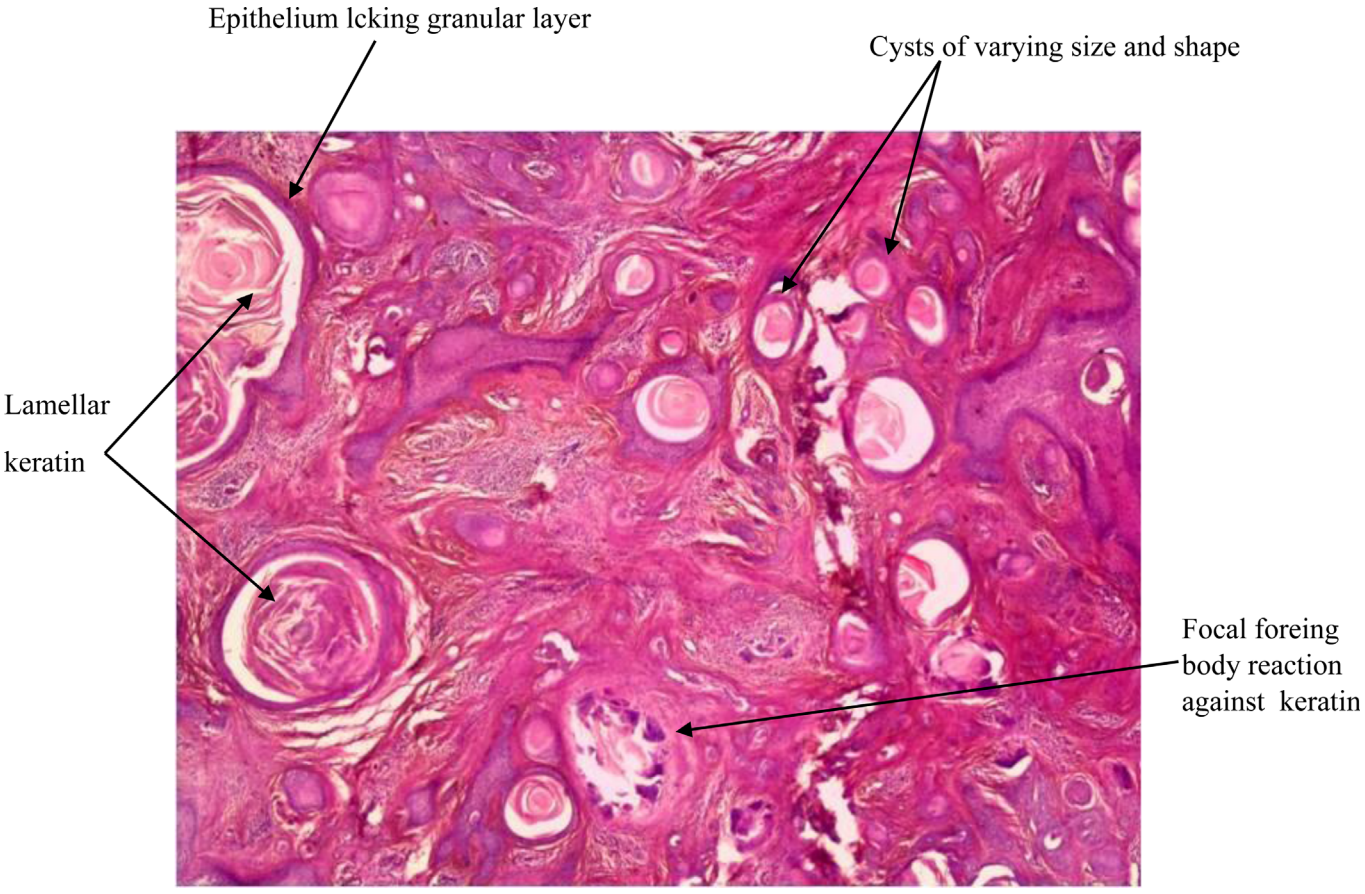

Fig. 3. Histological section following hematoxylin and eosin (H&E) staining(DrDerdour).

In the current WHO classification (5th edition, 2022), keratocystoma has been formally recognized as a distinct salivary gland tumor entity [6]. It was previously listed as a differential diagnosis of squamous cell carcinoma of the salivary gland by the WHO working group in 2005(3rd edition, 2005) [7].

Histologically, keratocystomas consist of multiple cystic spaces lined by squamous epithelium without cytologic atypia, necrosis, or invasive features. The absence of a granular cell layer and the presence of keratin lamellae within cystic spaces are distinguishing features. The epithelium is non-malignant, and there is no perineural or vascular invasion. Differential diagnosis includes: Squamous cell carcinoma (primary or metastatic), Mucoepidermoid carcinoma, Necrotizing sialometaplasia, Metaplastic Warthin tumor. The absence of cytologic atypia, necrosis, and invasive behavior supports a benign diagnosis [6].

Immunohistochemical staining typically shows positivity for AE 1/AE 3 and CK 5/6, and negativity for CK8/18, S100, and calponin [8]. Recent genetic studies have identified RUNX2 gene rearrangements in several cases, with an IRF2BP2::RUNX2 fusion found in most of them [9]. This genetic marker has not been found in other salivary gland tumors and may support keratocystoma's classification as a distinct neoplastic entity. Our case is histologically consistent with previously reported features, including keratin-filled cysts lacking a granular layer, but uniquely includes multinucleated Müller-type giant cells.

Conclusion

Keratocystoma is a rare, benign neoplasm of salivary gland origin, now recognized in the WHO 5th edition classification as a distinct tumor entity [6]. While most documented cases have arisen in the parotid gland, this report documents only the second known occurrence in the palatal minor salivary glands, thereby expanding the known anatomic spectrum of this neoplasm. Clinicians and pathologists should include keratocystoma in the differential diagnosis of well-demarcated, cystic palatal lesions, especially those lined by squamous epithelium with keratin accumulation and lacking signs of malignancy. Our case not only reinforces previously described histologic features but also introduces the observation of Müller-type giant cells, potentially enriching the morphologic understanding of keratocystoma. The absence of cytologic atypia and the favorable post-surgical course underscore the tumor's benign behavior. As more cases are reported, molecular testing may further validate RUNX2 rearrangements as a diagnostic hallmark and refine our understanding of its pathogenesis. Future studies should aim to clarify the biological behavior, recurrence rates, and potential genetic underpinnings of keratocystoma, particularly in minor salivary gland sites. Increased awareness and reporting will be essential to ensure accurate diagnosis and optimal patient care in cases of this rare tumor.

Funding

This research did not receive any specific funding.

Conflicts of interest

The authors declare no conflicts of interest in regards to this article.

Data availability statement

The research data is included in the article.

Informed consent

Consent has been obtained to use the patient's clinical photograph.

References

1. Seifert G, Jautzke G. Unusual choristoma of the parotid gland in a girl. Virchows Arch 1999;434:355–359.
2. Nagao T, Serizawa H, Iwaya K, *et al.* Keratocystoma of the parotid gland: a report of two cases of an unusual pathologic entity. Mod Pathol 2002;15:1005–1010.
3. Lépine C. Tumeurs des glandes salivaires : OMS 2022 et au-delà. Ann Pathol 2024;44:175–182.
4. Ahuja G, Aulakh KK, Sethi E, *et al.* Keratocystoma of minor salivary gland of the palate. Saint Int Dent J 2016;2:21–23.
5. Kokubun K, Chujo T, Yamamoto K, *et al.* Intraoral minor salivary gland tumors: a retrospective, clinicopathologic, single center study of 432 cases in Japan and a comparison with epidemiological data. Head Neck Pathol 2023;17:739–750.
6. Skalova A, Hyrcza MD, Leivo I. Update from the 5th edition of the WHO classification of head and neck tumors: salivary glands. Head Neck Pathol 2022;16:40–53.
7. Lewis JE, Olsen KD. Squamous cell carcinoma. In: Barnesy L, Eveson JW, Reichart P, Sidransky D, Eds. WHO Classification of Tumours. Pathology and Genetics of Head and Neck Tumours. Lyon: IARC Press; 2005:245–246.
8. Wolk RA, Cipriani NA. Update of newly recognized salivary gland neoplasms: molecular and immunohistochemical findings and clinical importance. Histopathology 2025;86: 183–198.
9. Bishop JA, Nakaguro M, Urano M, *et al.* Keratocystoma: a distinctive salivary gland neoplasm characterized by RUNX2 rearrangements. Am J Surg Pathol 2024;48:317–328.

Cite this article as: Benaouf S, Kraloua S, Larbi M, Khebichat MEA, Tabeti-Bentahar CF. 2025. Keratocystoma of palatal minor salivary glands: a case report. *J Oral Med Oral Surg*. 31: 35. https://doi.org/10.1051/mbcb/2025038