Benign myoepithelioma of the soft palate: an unusual clinical entity

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SUMMARY

Myoepitheliomas are rare benign tumours that affect the exocrine glands and are sporadically located in the salivary glands. The most common location of myoepithelioma in the oral cavity is the parotid gland and it is seldom encountered in the palate. The diagnosis of this entity is challenging since its clinical presentation may resemble those of more common neoplasms, rendering a complex histopathological diagnosis. The aim of the present case report is to describe an unusual case of myoepithelioma of the soft palate in a male patient, which developed as an asymptomatic, slowly growing mass. The tumour was assessed with histopathological examination and the diagnosis was verified via immunohistochemistry. Finally, the treatment included surgical resection of the tumour and no signs of recurrence were noted 2.5 years after the surgical procedure. Early diagnosis and treatment plays an important role in the prognosis of this pathological entity.

BACKGROUND

Myoepithelioma is a rare benign tumour encountered in the salivary glands and mainly constitutes ectoderm-derived contractile cells that act as smooth muscle cells and have a role in gland secretion. Myoepithelial cells in the salivary glands are located between the basal lamina and the acinar and ductal cells.[1] The tumour was formerly regarded as a pleomorphic adenoma subtype, and thereafter was referred to as 'monomorphic adenoma', 'myoepithelial adenoma', 'myoepithelial cell tumor', 'adenomyoepithelioma' and 'parotid clear cell adenoma', among other names. However, the term 'myoepithelioma' was first used by Sheldon in 1943 to describe the pathology.[1 2] In 1991, the WHO recognised the tumour as an independent entity and is currently categorised as a benign tumour of the salivary glands in the Classification of Head and Neck Tumors of 2017.[1 2]

Myoepithelioma accounts for 1%–1.5% of all benign and malignant tumours that affect the salivary glands and represents 2.2% and 5.7% of all benign major and minor salivary gland tumours, respectively.[2 3] This neoplasm will primarily affect the parotid gland in approximately 40% of cases, followed by minor salivary glands in 21%, with the most common sites of occurrence being the hard and soft palate.[2 4] Neoplastic myoepithelial cell tumours can be found in the vestibular mucosa, labial mucosa, nasopharynx, nasal septum, breast, sweat glands, lacrimal glands, trachea, larynx, lung, oesophagus, retroperitoneum and prostate gland. The pancreas would be one exception.[4]

Myoepitheliomas frequently affect patients between the fourth and fifth decades of life, without gender predominance.[5] Clinically, it presents as a slowly growing, asymptomatic, solid mass in major or minor salivary glands.[2]

CASE PRESENTATION

A 56-year-old man with a history of controlled type 2 diabetes mellitus presented to the oral and maxillofacial department with a large mass in the soft palate and a foreign body sensation. He stated that the mass is painless and that it has been slowly growing for 15 years. No dysphagia, dyspnoea, odynophagia, dysphonia or weight loss was reported by the patient. Physical intraoral examination revealed a firm, mobile, non-fluctuating and well-defined tumour of the soft palate with an approximate diameter of 2×2 cm, with a colour similar to the adjacent mucosa and with a smooth surface (figure 1). There was no pathological cervical lymphadenopathy reported and the remainder of the general examination revealed no alterations.

INVESTIGATIONS

Contrast-enhanced CT of the head and neck region revealed an isodense image corresponding to the adjacent soft tissue with an approximate size of 27×30×24 mm. A homogenous, non-contrast-enhancing, encapsulated, well-defined tumour was located in the soft palate and extending to the oropharynx, without infiltrating into other structures. No evidence of bone erosion or cervical lymphadenopathy was observed (figure 2). Findings from tomographic imaging suggested that this could be a benign tumour, based on the features previously described. As a second diagnostic tool, it was decided to proceed with fine needle aspiration biopsy (FNAB); however, the results from the pathological report were inconclusive of diagnosis. In this scenario, the case was reassessed considering the clinical and imaging characteristics of the tumour, such as non-tender, mobile mass, with irregular borders, smooth surface, no changes in mucosal colour, long evolution, absence of lymphadenopathy and non-adherence to deep structures. The latter characteristics suggest that this could be a benign lesion. The final treatment plan was decided, which included excisional biopsy of the tumour.

Figure 1 Mobile, solid and well-defined tumour located on the right side of the soft palate.

DIFFERENTIAL DIAGNOSIS

At first, the differential diagnosis included pleomorphic adenoma based on the diagnostic information previously acquired, since it is the most common tumour affecting the salivary glands (60%) compared with myoepithelioma (1%).[5] The second differential diagnosis focused on benign lesions arising from the connective tissues (neural, muscle and adipose tissue), given the anatomical location and clinical features of the tumour. Malignant transformation or infection was not considered because the tumour was asymptomatic, with a slow-growing rate, and there was no reported lymphadenopathy.

TREATMENT

The tumour was removed via intraoral approach under a balanced general anaesthesia, without rupture of the fibrous capsule. The specimen was assessed by the pathology department (figure 3), and recovery of the patient after the surgical procedure was uneventfu

OUTCOME AND FOLLOW-UP

Histopathological examination rendered a diagnosis of myoepithelioma of the minor salivary glands without infiltration to the capsule or to the adjacent adipose tissue. No signs of necrosis or mitotic activity were observed, and the diagnosis was confirmed with immunohistochemical reactions to positive markers (figures 4 and 5 and table 1). A regular follow-up was conducted and no signs of recurrence were observed at 30-month evaluation (figure 6).

DISCUSSION

Myoepithelioma is a benign tumour that may be found in nearly all exocrine gland tissues. This neoplasm frequently arises in the parotid gland, followed by the minor salivary glands, where the palate is the most frequent location, accounting for 93% of the cases found intraorally.[2 4]

CT imaging may reveal a well-demarcated, homogeneous or heterogeneous, smooth or lobulated mass, and in some instances adjacent bone erosion may be observed.[4 6] CT imaging allows for assessment of the relationship between the salivary gland tumours and the gland parenchyma and the adjacent soft tissues. Moreover, this evaluation reveals changes in the adjacent bone and the extension of the neoplasm to other anatomical spaces, as well as the presence of non-palpable lymphadenopathy. The use of contrast-enhanced CT is advocated since contrast media uptake is different in normal vascular tissues compared with that of a tumour, which demonstrates an increased blood supply. The latter allows distinction of the adjacent tissues.[7] In the present case a contrast-enhanced CT revealed a well-demarcated, heterogeneous mass, without adjacent bone erosion.

A useful imaging modality is contrast-enhanced MRI. Myoepithelioma palatal lesions show hypointensity on T1-weighted images and hyperintensity on T2-weighted images. MRI is the best imaging modality for evaluation of salivary gland tumours, particularly for malignant palatal tumours. Contrast MRI accurately locates the tumour and its extension into the adjacent soft tissue. Malignant palatal tumours may demonstrate perineural spread along the greater and lesser palatine nerves, followed by extension to the pterygopalatine fossa and cavernous sinus.[6] In the presence of a malignant tumour of the salivary glands, MRI should be the first-choice imaging modality.

Differential diagnosis of myoepitheliomas should include benign tumours (pleomorphic adenoma, schwannoma, neurofibroma, leiomyoma, benign fibrous histiocytoma and extramedullary plasmacytoma), malignant salivary gland neoplasms (rhabdomyosarcoma, mucoepidermoid and myoepithelial

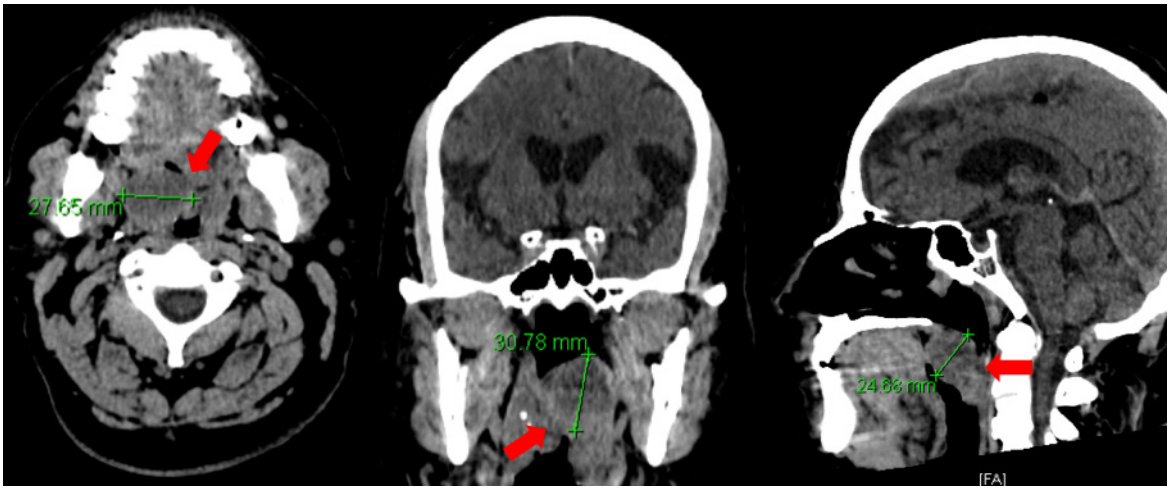

Figure 2 Contrast-enhanced CT images showing a 27×30×24 cm tumour (arrows) on the right side of the soft palate. An isodense, homogeneous and non-contrast-enhancing tumour is observed, extending from the soft palate to the oropharynx.

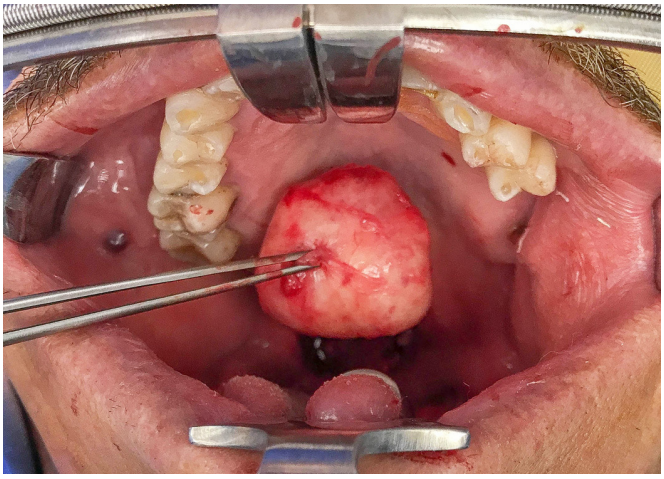

Figure 3 Surgical excision of the lesion. The specimen was covered by a fibrous capsule.

carcinoma), as well as mucocoele, abscess, and odontogenic cysts and tumours.[3 5]

In the presence of salivary gland tumours, FNAB is recommended for cytological evaluation prior to obtaining an incisional biopsy. The aims of FNAB are to distinguish between neoplastic and non-neoplastic lesions, determine neoplasm lineage, and distinguish between benign, malignant and metastatic tumours.The sensitivity of this diagnostic tool ranges from 57.9% to 86% and results in a specificity of 98.8%. Diagnosis of benign or malignant entity can be made in 98% and between 60% and 70% of cases, respectively.[8] Occasionally, cytological evaluation may produce a non-diagnostic result (Milan system), which generally occurs as a result of lack of cellularity or artefacts that affect interpretability.[8 9] Only two case reports in the literature have described the diagnosis through cytopathological findings using FNAB.[10 11]

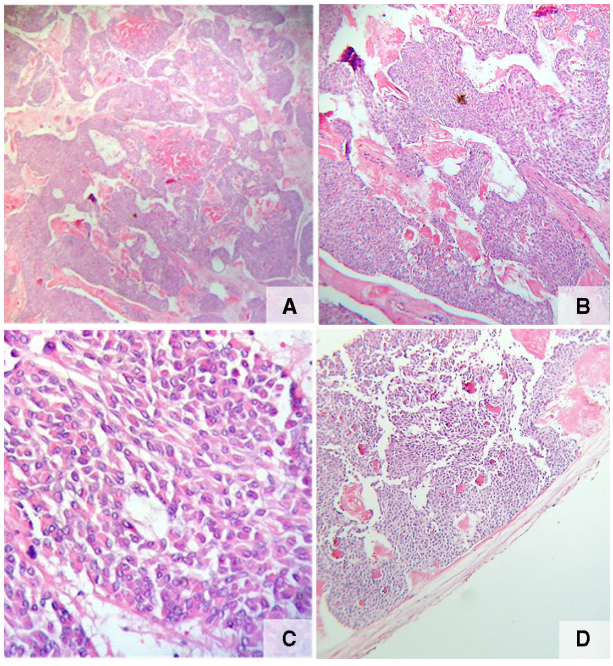

Figure 4 H&E stain microphotographs. (A, B) Note the neoplastic organisation in niduses separated by hyaline stroma (×4). (C) Plasmacytoid cells mixed with spindle cells (×40). (D) The tumour reveals well-defined, regular borders, as well as a fibrous capsule (×10).

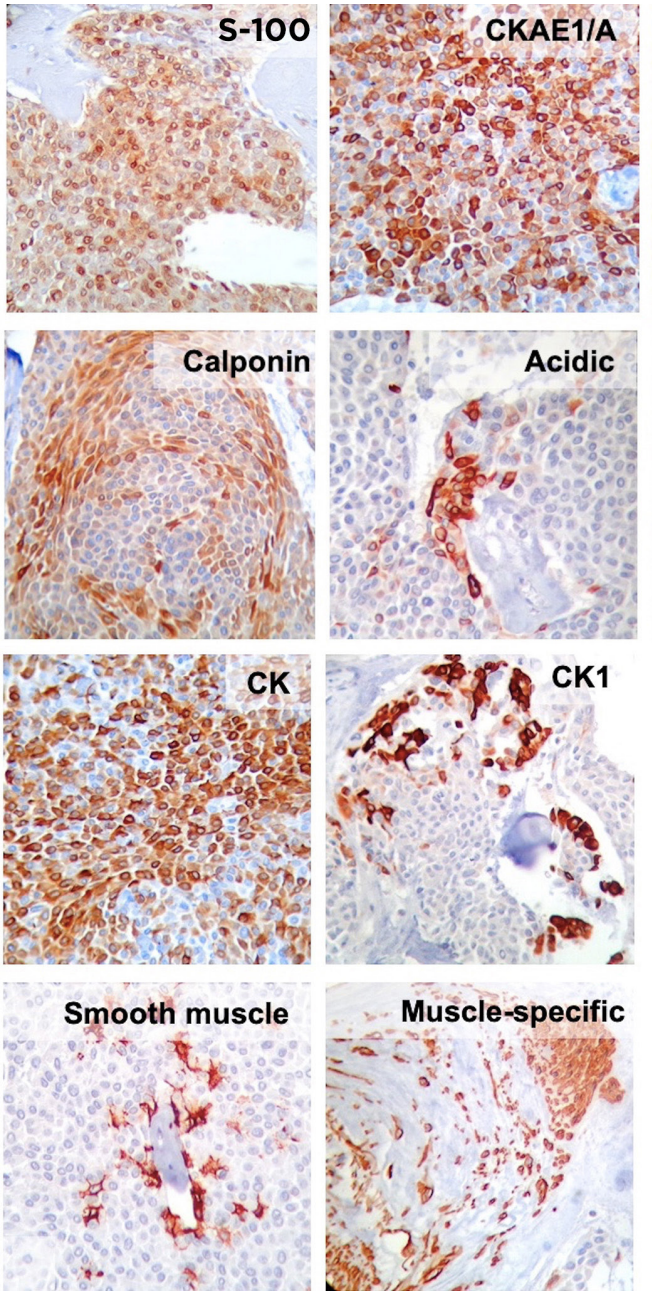

Figure 5 Photomicrography corresponding to immunohistochemical findings. The tumour cells reveal an intense positivity in more than 90% of the cell population for S-100, CKAE1/AE3, CK7 and calponin. On the other hand, glial fibrillar acidic protein shows an intense positivity in 15% of cells. CK14, smooth muscle actin and muscle-specific actin reveal focal positive staining in 10% of the cell population (×10 magnification).

Myoepithelioma may present different histological, immunohistochemical and molecular characteristics, rendering diagnostic and surgical challenges. Clinically, it presents as a solid, irregular, well-demarcated and slowly growing mass or tumour that is non-tender and with long evolution.[5] Once a biopsy is obtained and a macroscopic analysis performed, a solid tumour with a distinct peripheric border may be observed. The nucleus presents a white-yellow colour. The tumour becomes semitranslucid when the myxoid extracellular matrix is abundant.[1]

Table 1 Panel of immunohistochemical markers showing positive and negative staining in the tumour tissue

Immunoreactive	+/–	Expression level
S-100	+	Intense
CKAE1/AE3	+	Intense
CK7	+	Intense
Calponin	+	Intense
Glial fibrillar acidic protein	+	Heterogeneous
Smooth muscle actin	+	Heterogeneous
Muscle-specific actin	+	Weak
CK14	+	Weak
p63	–	Negative

Clinical differentiation between benign and malignant tumours that affect the salivary glands has been described in the literature. Some of the clinical characteristics that may predict malignant behaviour of the tumour located in the palate have been outlined, which include pain, irregular surface, changes in the colour of the mucosa and ulceration. However, these are not pathognomonic of malignancy and each case must be individually assessed.[12] In the present case, the tumour lacked the latter features related to malignancy, presuming that it could be a benign lesion.

Regular and well-defined borders, presence of a fibrous membrane (sometimes incomplete when the lesion originates in a minor salivary gland) and a parenchyma with a diverse histological profile are some of the microscopic features found in myoepitheliomas. Depending on the specific cell differentiation stage, several cell subtypes may be distinguished. Epithelioid or epithelial cells account for 45% of the main body of the tumour, followed by spindle cells in 32.5%, plasmacytoid cells in 7.5%, and hyaline and clear cells in 2.5%. One specific cell type may be predominant, or a combination of different cell types may be present. It has been estimated that 12.5% of tumours have a main body with a mixed cell type.[1] Moreover, the morphological architecture can exhibit the following growth patterns: solid, non-myxoid pattern (60% of myoepithelioma tumours) with cells growing densely and accompanied by a fibrous stroma; a myxoid pattern with tumour cells growing in an insular, trabecular and sporadic fashion in an abundant myxoid matrix; a reticular pattern with a trabecular structure of tumour cells on a myxoid or hyaline matrix; and a mixture of the three growth patterns.[1 13] Myoepitheliomas are distinguished from pleomorphic adenoma, which is regarded as the main differential diagnosis, by lack of both ductal structures and chondroid or myxochondroid matrix.[13] The present case exhibited a reticular pattern with a hyaline/myxoid matrix and abundant plasmacytoid cells (90%), intermixed with a scarce number of spindle cells (figure 6).

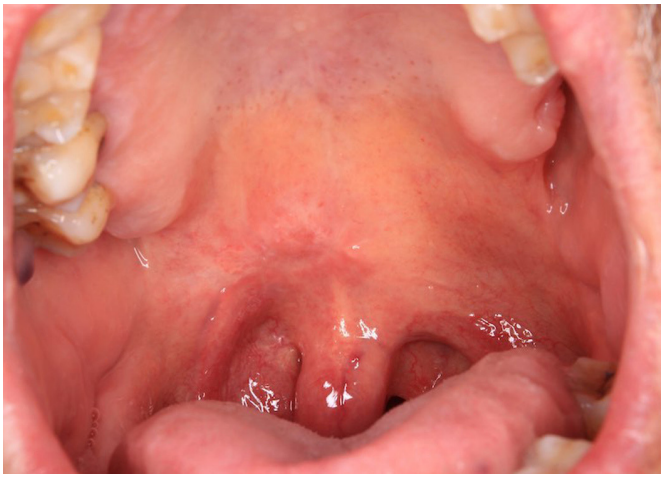

Figure 6 Clinical image revealing postsurgical condition at 6 months.

It should be pointed out that, given its benign nature, the tumour lacks infiltrative borders, perineural infiltration, vascular lymphatic permeation, necrosis, nuclear pleomorphism and/or atypia, and mitotic activity. The presence of the aforementioned features should warn the oral pathologist on the possibility of a myoepithelial carcinoma (a malignant and infrequent variant of myoepithelioma).[14]

Immunohistochemical analysis can aid in the diagnosis, with immunoreactivity or positivity to CK7 CK14, CKAE1/AE3 (90%–100%), S-100 (72%–100%) and p63 (60%), among other markers such as GFAP (27%–54%) and calponin (86%–100%). Alpha smooth muscle actin and calponin positivity may vary and often occurs in the spindle cell component. On the other hand, the latter proteins may be negative for plasmacytoid component. Desmin is primarily negative (0%–20%).[4 10]Genetic rearrangements in the EWSR1 gene have been reported in 45% of cases.[10] The present case demonstrated immunohistochemical panel with S-100 (100%), CKAE1/AE3 (90%), CK7 (90%), calponin (90%), weak positive focal staining of CK14, positive heterogeneous glial fibrillar acidic protein, smooth muscle actin positivity in the spindle cell component, weak positive focal staining of muscle-specific actin and negative p63. The diagnosis was confirmed based on these observations (figure 4).

The treatment of choice is surgical excision. Recurrence is unusual and is generally associated with incomplete removal of the lesion. Some authors report a recurrence rate of 15%–18%, while other studies demonstrated that only 1 out of 16 myoepitheliomas recurred over a 7-year period. Moreover, recurrence of this tumour may arise as a malignant component, which has been attributed to the overexpression of receptors and p53 mutations.[4]

Learning points

- Benign tumours located in the salivary glands represent a diagnostic and therapeutic challenge for clinicians, and therefore a thorough understanding of the pathologies that may affect these structures is paramount.
- Although myoepithelioma is an uncommon tumour, it must be considered as a differential diagnosis of tumours that derive from salivary glands, especially minor salivary glands and particularly those located in the palate, which is the most affected site in the oral cavity by benign tumours and the second most affected by myoepithelioma.
- Complementary imaging and cytological studies will guide the diagnosis and treatment plan and must be performed in the presence of tumours in the salivary glands.
- Establishment of a specific clinical, imaging and cytological diagnosis of myoepithelioma represents a challenge.
- Treatment for this pathology is like other benign lesions that affect minor salivary glands, namely surgical excision, and the final diagnosis is determined by histopathological examination.

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Contributors MGM participated in the conception, planning and design of the clinical case, acquisition of data, taking clinical photographs, bibliographic search and elaboration of the manuscript. AJ participated in the conception, planning and design of the clinical case, acquisition and interpretation of data, and critical revision of the manuscript. FS participated in the conception and elaboration of the manuscript, acquisition and interpretation of data, and search for bibliographic references that support the article and in its critical review. GMGM participated in the data analysis, issued the pathological diagnosis of the clinical case and critical review of the manuscript. All authors approved the final version of the manuscript.

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