

Using the Le Fort I approach in removing a palatal mucoepidermoid carcinoma



Xander Jackers^{a,*}, Robin Snel^a, Michel Bila^a, Constantinus Politis^{a,b}

^a Department of Imaging and Pathology & Department of Maxillofacial Surgery, University Hospitals Leuven, Leuven, Belgium

^b OMFS-IMPATh Research Group, Catholic University Leuven, Leuven, Belgium

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ABSTRACT

Mucoepidermoid carcinomas are a relatively uncommon type of malignancy of the head and neck region. Traditionally, they are classified into low-, intermediate- and high-grade lesions by the WHO which reflect clinical behaviour. However, this classification remains a point of discussion with alternative classifications at hand. Surgery is the preferred treatment and is combined with radiotherapy in high grade lesions or specific situations.

In this case report, a case of a mucoepidermoid carcinoma (MEC) of the right hard palate in a 36-year old patient is presented. The lesion, however, was first diagnosed as a benign cystic lesion after which a mucoepidermoid carcinoma was found within on histopathological examination. The malignancy was surgically removed including the underlying palatal bone and the greater palatine canal guided by a Le Fort I approach. This approach offers direct visualisation of the resection margins and the canalis pterygopalatina. Reconstruction was carried out by using an ipsilateral buccal fat pad flap, leucocyte and platelet rich fibrin membranes (L-PRF). The remaining defect was covered by a palatal cover plate afterwards. In this report, we add detailed illustrations of the surgical approach in which the important anatomical landmarks are pictured.

1. Introduction

Salivary gland carcinomas are a relative uncommon category of tumours in the head & neck region. They represent 5% of the head and neck neoplasms. Approximately 10–15% of the salivary gland neoplasms occur in the minor salivary glands as was the case in this patient. Eighty percent of the minor salivary gland tumours are malignant [1]. The most common malignant salivary gland tumour is the mucoepidermoid carcinoma (MEC), closely followed by the adenoid cystic tumour [2]. They comprise around 10% of malignant salivary tumours [3].

MEC of the minor salivary glands can arise in the palate (most prevalent), the retromolar area, the floor of the mouth, the buccal mucosa, the lips and the tongue.

Rarely, they arise at laryngeal, nasal, paranasal, lacrimal, tracheal and pulmonary sites [2].

In this case, we present a MEC of the palatal minor salivary glands which is managed by surgery alone. It was carried out through a Le Fort I approach. A Le Fort I approach with downfracture simplifies the surgical procedure by improving the visualisation of the entire tumour and the nearby structures, such as the descending palatine artery and pterygopalatine nerve. In this manner, a complete tumour resection is guaranteed and the potential complications are reduced [4]. This report adds to the current literature by explaining and

* Corresponding author. Department of Oral and maxillofacial Surgery, University Hospitals Leuven, Kapucijnenvoer 33, 3000 Leuven, Belgium.
E-mail address: xander.jackers@live.com (X. Jackers).

picturing this type of approach in a comprehensive way.

2. Report of case

A 36-year-old female patient presented at the department to the Department of Oral and Maxillofacial Surgery of the University Hospitals of Leuven, (Leuven, Belgium) with a 5-year history of a palatal swelling on the right side. The lesion was characterized by a slow and insidious growth over the last few months. There was no associated pain or discomfort noted. Her past medical, social and familial history were unremarkable. She did not take any medications. Furthermore, the patient had no other complaints. On clinical examination, a well-defined and oval shaped lesion was described at the right side of the palatine raphe (hard palate). The anterior border was at the level of the first premolar. Posteriorly, it reached to the level of the first molar and no mobility or displacement was noticed. The lesion did not cross the midline of the palate (Fig. 1). Dental examination showed positive sensitivity to dry ice testing of the teeth in the upper jaw. The lesion had a cystic, bluish appearance in the middle with slightly erythematous borders. There were no ulcerations visible. The surface was smooth, and the lesion was firm on palpation. An area of hypoesthesia was noted posteriorly of the lesion. There was no tenderness or bleeding on palpation.

A panoramic radiograph (Vistapano, Durr Dental, Bissingenheim, Germany) was done which was unremarkable. An additional cone-beam computed tomographic (CBCT) scan (90 kV, 12 mA; Scanora 3D, Soredex, Tuusula, Finland) showed an expansive, nodular soft tissue lesion located at the posterior area of the hard palate with scalloping of the underlying bone (see Fig. 2).

An incisional biopsy of the cystic lesion was performed under local anaesthesia and histopathological investigation was suggestive of a mucocele. The entire lesion with underlying periosteum was excised afterwards under local anaesthesia because of its tendency to recur and the specimen was sent for histopathological investigation. The second pathology report, however, showed a mucoepidermoid carcinoma. A FISH-technique for MAML-translocation was positive and confirmed the diagnosis of a mucoepidermoid carcinoma.

Pre-operative oncologic work-up was carried out after resection. A computed tomography (CT) scan of the head and neck region showed the status after mucosal resection on the hard palate. Infiltration of the soft tissues in the greater palatine canal on the right side was apparent but non-specific. Differentiating sequela after the resection from tumour invasion in the canal was not possible.

Because of inability to exclude tumour invasion and the positive margins on histopathology, a broader resection was carried out through a Le Fort I approach which included the bone underlying the lesion. Firstly, an access window to the right maxillary sinus was done and the sinus mucosa was lifted. Subsequently, osteosynthesis plates and screws (KLS Martin, Freiburg, Germany) were used to secure the exact position of the maxilla. Access to the surgical site was then achieved by the standard Le Fort I osteotomy. A wider mucosal excision was performed around the previous defect. The greater palatine canal was marked circumferentially. The greater palatine artery was then clipped. Afterwards, the palatine bone underlying the lesion was removed including the greater palatine canal and its contents by means of piezotomy (Piezosurgery 3, Mectron, Carasco, Italy).

Detailed illustrations of the surgical procedures show the Le Fort I osteotomy and the superior view of the palate with important anatomical structures (Fig. 3). A sagittal view is also provided.

The right buccal fat flap was prepared and attached to the left palate for reconstruction to support the bottom of the sinus mucosa. The former osteosynthesis plates were used to reduce the osteotomy to its original position, L-PRF (Leucocyte and Platelet Rich Fibrin) membranes were applied to enhance wound healing [5].

Immediately following closure of the wounds, a palatal cover plate (constructed pre-operatively) was placed over the defect which provided adequate prosthetic reconstruction and excellent functional repair post-operatively. Fig. 4 shows a posterior part of the resected specimen (see Fig. 4).

Post-operative histopathological assessment of the resection specimen showed no remaining tumour, nor invasion of the bony palate. Fig. 5 (histopathological) shows the tumour and its characteristics.

Regular follow-up was done at 1 week, 3 weeks, 1 month and showed a favourable evolution of the wound. At week 1 and 3 post-



Fig. 1. Photograph at initial examination showing a well-defined and oval shaped lesion which was described at the right side of the palatine raphe (hard palate).

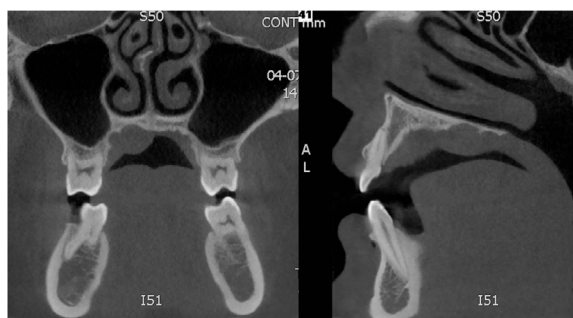


Fig. 2. Expansive, nodular soft tissue lesion located at the posterior area of the hard palate with scalloping of the underlying bone. **Left:** Coronal view, **right:** Sagittal view.

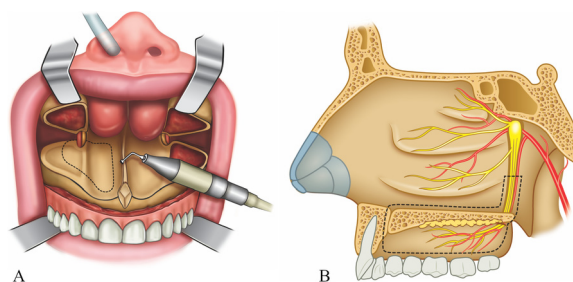


Fig. 3. (A) Le Fort I downfracture with superior view on the hard palate. The dotted line marks the resection margins which includes the lower part of the pterygopalatine canal and its contents (greater palatine nerve and artery). (B) sagittal view on the resection margins with part of the pterygopalatine canal.

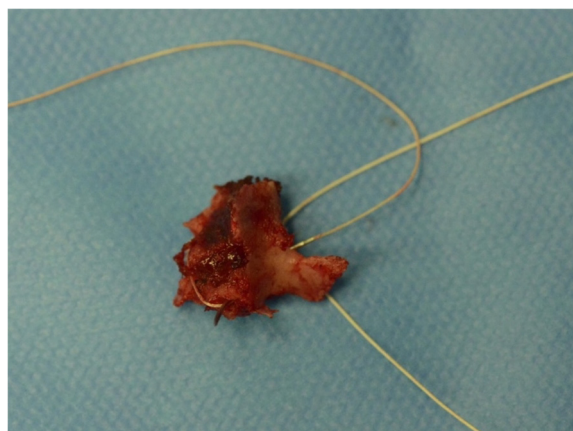


Fig. 4. Resection specimen of the hard palate after removal through a Le Fort 1 approach. Orientation of the specimen with sutures. The needle is passed through the greater palatine canal.

operatively, L-PRF was again applied in the outpatient's department to enhance healing of the palatal defect.

3. Discussion

As mentioned before, MEC is a relatively uncommon malignancy in the head and neck region. It is, however, the most common type of malignant tumour of the minor salivary glands.

They are mostly seen in patient in their fourth to seventh decade but can occur at any age. Women tend to have a slight predilection [6].

Clinically, they often present as a painless, rubbery or soft mass with variable fixation. Intraoral tumours can be blue-red tinged resembling a mucocele or vascular tumour. The former was the case in this patient. Erosion of underlying cortical bone can be present. Other possible symptoms include ulceration, pain, fluctuation, discoloration and nerve palsy [7].

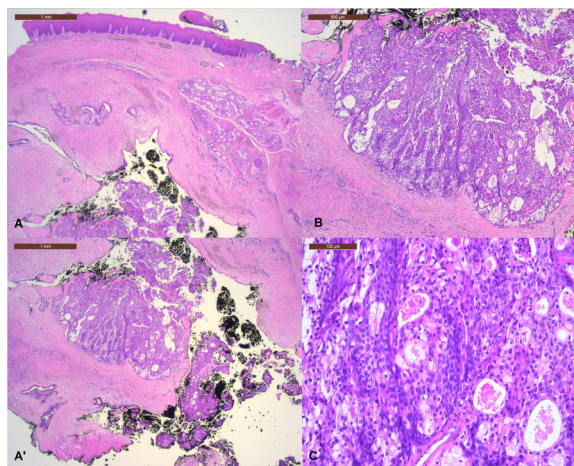


Fig. 5. A (H&E staining, 25 $\times$ magnification). The tumour is seen at the bottom. Black ink is used to mark the resection margins. This picture depicts the relationship of the mucoepidermoid carcinoma with the oral mucosa (upper part of the picture, stratified squamous epithelium); A' (visualisation of the entire tumour, 50 $\times$ magnification) Visualisation of the tumour. Dilated glandular tissue with a cystic appearance is seen. This is surrounded by an intense eosinophilic stroma (i.e. hyalinisation of the stroma). Nests of non-encapsulated proliferating intermediate cells and clear cells are close to the cystic structures. In these nests, lumina with eosinophilic material inside are noted; B (H&E staining, 200 $\times$ magnification) Individual cells can be distinguished on this level of magnification. Their cytoplasm ranges from eosinophilic to clear. Only a few cells contain mucus. They also have clear borders. The nuclei are vesicular with mainly fine chromatin. Anisokaryosis is rare and increased mitotic activity is not apparent.

3.1. Diagnosis

If opposed to a swelling of the palate, imaging is the next step in the diagnostic process. Imaging is preferably performed by means of computed tomography (CT) or magnetic resonance imaging (MRI). These are mandatory in estimating the anatomic disease extent [1]. A CT-scan is preferred for assessment of bony invasion whereas an MRI is superior in evaluating soft tissue. Positive emission computed tomography (PET-CT) is a valuable alternative, especially in loco-regionally advanced tumours according to a review in 2015 [7].

Histopathological investigation remains the golden standard for diagnosis [7]. Vander Poorten et al. state that an incisional/punch biopsy is preferred over fine needle aspiration biopsy for diagnosis of salivary gland tumours and, importantly, does not interfere with final surgery [1].

The literature contains different histological grading systems for MEC. The Armed Forces Institute of Pathology (AFIP) presented a first classification which is used by the WHO. Tumours are traditionally graded into 3 different histopathological groups (low-, intermediate- and high-grade variant). Brandwein et al. state that the AFIP criteria tend to downgrade MEC in discrepant cases and modified the scoring system to enhance the predictive value [6]. A third grading system is the Modified Healey system. Intermediate grade tumours have the most variability between grading systems and subsequently demonstrate controversy in management [8].

3.2. Management & Le Fort I approach

Surgery with the goal of acquiring complete free margins is the primary treatment of MEC. In small, low grade MECs, local wide resection is considered curative. In locally advanced disease, more elaborate surgical planning is frequently necessary (maxillectomy, infratemporal fossa approach, hard palate resection etc.) [7]. Early surgical management of palatal salivary minor gland tumours included wide local excision and enucleation if benign and hemimaxillectomy for malignant tumours. Recently, more tissue-sparing techniques like alveolectomy, palatectomy and infrastructure maxillectomy are put forward [9]. Some authors propose to perform exclusively bony resection of the palate if radiological and/or perioperative findings of bony invasion are present. If not present, wide mucosal resection is sufficient [10]. We performed bony resection because of the positive deep margin on the first mucosal resection specimen (including periosteum) and uncertainty whether the greater palatine canal was infiltrated with tumour tissue on radiology.

Von Langenbeck and Cheever provide the first definitions of the Le Fort I osteotomy for removal of nasopharyngeal tumours in mid-19th century. The Le Fort I osteotomy is well known as an approach for tumours of the midface, nasopharynx and skull base [11]. The literature is not as extensive on the use of this approach specifically for intra-oral palatal carcinomas however. De Cevallos et al. reported a case in which this approach was used for resection of a palatal tumour [4]. One report did also use this approach for a pleomorphic adenoma of the palate and reported a very satisfactory post-operative functional as well as esthetical result [12]. We agree with the statement that this approach allows direct visualisation, increases safety and minimizing the possibility of recurrence [12]. The Le Fort I approach yields improvement of the visualisation of the resection site and the related structures such as the descending palatine artery and nerve [4].

Concomitant radiation therapy is advised in high grade tumours, unclear margins, perineural invasion, high tumour staging and patients with positive lymph nodes [2,7].

Neck dissection is indicated if there is clinical or radiologic evidence of regional metastasis [1]. Elective dissection, however, remains

a difficult decision. It is accepted that elective neck dissection should be performed in high-grade tumours [1,7].

Reconstruction of malignancies used to be performed by means of local flaps or were obturated. Recent changes advocate primary reconstruction of the palatomaxillary defect. Newer techniques for reconstruction are local and regional pedicled flaps (tongue flaps, uvular flaps, the palatal island flap, buccal flaps, temporalis myofascial flaps, submental flaps) and free tissue transfer. Reconstruction of the palate is, however, complex due to the complex functions of the palate during swallowing and speech [9].

3.3. Prognosis

Low-grade MECs have a good prognosis whereas high-grade MECs have a worse prognosis because of their more aggressive behaviour. Investigations show that prognosis of intermediate-grade MECs do not differ significantly from low-grade tumours [7,13,14]. Low- and intermediate grade tumours combined have a 5-years disease specific survival of 97.8% versus 67.0% for the high-grade tumours [14].

Negative prognostic factors include primarily high histological grade and advanced clinical stage. Other negative factors are positive surgical margins, perineural and vascular involvement, and lymph node metastasis [15].

In this case, investigations for MAML2 gene arrangement were carried out and came back positive. Luk et al. found that this test has a sensitivity of 82% and a specificity of 100%. Conclusively, this test is a useful tool in diagnosing the tumour. Whether the MAML2 gene rearrangement test also carries prognostic value is not clear yet [16].

4. Conclusion

Palatal MEC is a relatively uncommon malignancy of the head & neck area. It should be diagnosed with an incisional/punch biopsy after imaging by means of CT and/or MRI to assess for tumour size and expansion. MECs are mostly graded into low, intermediate and high-grade variants although different authors propose other grading systems. The high-grade variant is particularly aggressive and had an unfavourable prognosis. Low and intermediate graded tumours behave less aggressive and can be managed solely by wide local resection. Radiation therapy is added in particular circumstances as discussed earlier. Important prognostic factors are primarily histologic grade and tumour staging.

This is one of the few reported cases of a salivary gland tumour of the palate managed by a Le Fort 1 approach with subsequent resection of the right palate. If opposed to a palatal MEC or other carcinomas (adenoid cystic, squamous cell tumour ...), this approach offers valuable alternative to the classic *trans*-oral approach. We propose that this technique provides an excellent visualisation of the resection margins and visualizes important related structures such as the greater palatine vessel and greater palatine nerve with low morbidity and acceptable functional and aesthetic outcome.

Conflicts of interest

None.

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Informed consent

Informed consent for publication was obtained from the patient.

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