

CASE REPORT

Salivary glands: report of a rare case of myoepithelial carcinoma involving tongue base treated by CO₂ Laser

Ghiandole salivari: case report di un raro caso di carcinoma mioepiteliale della base della lingua trattato con il laser a CO₂

E. ESPOSITO, B. CASSIANO, F. CINQUEGRANI¹

ENT Dpt., "S. Maria Della Pietà" Hospital, Nola; ¹ ENT Dpt., "Vecchio Pellegrini" Hospital, Naples, Italy

SUMMARY

Epithelial-myoepithelial carcinoma is a rare malignant tumour of the salivary glands. Herewith, the clinical evolution, anatomic-pathological characteristics and treatment adopted are described in a case occurring in the minor salivary glands of the tongue base. To our knowledge, this location has not been previously described in the literature. In fact, due to the rarity of the epithelial-myoepithelial carcinoma, there is no uniformity of data in the literature and very different therapeutic strategies have been suggested. In this report, the possibility of applying conservative surgery using CO₂ Laser is proposed. Results obtained in the case described confirm that conservative surgery is feasible in this type of neoplastic lesion of the tongue.

KEY WORDS: Myoepithelial carcinoma • CO₂ Laser

RIASSUNTO

Il carcinoma mioepiteliale è una neoplasia che raramente interessa il tessuto ghiandolare. Gli Autori illustrano un caso di carcinoma mioepiteliale insorto nel tessuto salivare della base della lingua e trattato con successo mediante il laser a CO₂. Data la rarità della neoplasia, non sono descritti precedenti in letteratura né, conseguentemente, sono illustrati protocolli terapeutici. In conclusione, gli Autori dimostrano che la chirurgia conservativa è possibile per questo tipo di lesioni.

PAROLE CHIAVE: Carcinoma mioepiteliale • Laser a CO₂

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Introduction

Epithelial-myoepithelial carcinoma is a very rare low grade malignancy epithelial tumour, characterised by the presence of two well-differentiated epithelial cell populations: the myoepithelial cells, which appear greater, clear staining and peripherally arranged around the second population, the ductal cells, that are central and may be present in increasing amounts, but usually inferior in comparison to the myoepithelial cells ¹.

This tumour has an incidence of 1% of all tumours of the salivary glands and involves predominantly the major glands, in particular the parotid gland. Even if data in the literature are very different ^{2,3}, it can be argued that this tumor has a percentage of onset in the intra-oral minor salivary glands that varies between 10 and 15%.

Several unusual locations of this carcinoma have been described such as, for instance, the nasal cavity, the paranasal sinuses or the lacrimal glands ⁴⁻⁶ but, so far, onset in the minor salivary glands of the tongue base has never been described.

The observation of such an exceptional clinical case, is

of scientific interest not only on account of the unusual finding, but also because it gives rise to issues concerning the correct therapeutic strategy to apply. In fact, there was no possibility to refer to schemes or therapeutic protocols previously adopted for which the indicative percentages of recovery are usually reported.

Case report

In April 2000, the patient A.T., a 67-year-old female, went to an ENT specialist because she was suffering from dysphagia, right otodynia, a cough caused haemoptysis and feeling of a foreign body in her throat. The clinical examination revealed a neof ormation at the base of the tongue that was extended to the glossoepiglottic fossa and to the epiglottis (Fig. 1); on the basis of this finding it was recommended that she be hospitalised.

A few days later she entered the "St. Gennaro" Hospital of Naples in order for a biopsy specimen to be collected in the region of interest. Histologically the tumour showed a multilobular architecture, characterized by epithelial nests divided by fibrous septa. Epithelial cells showed a trabecu-

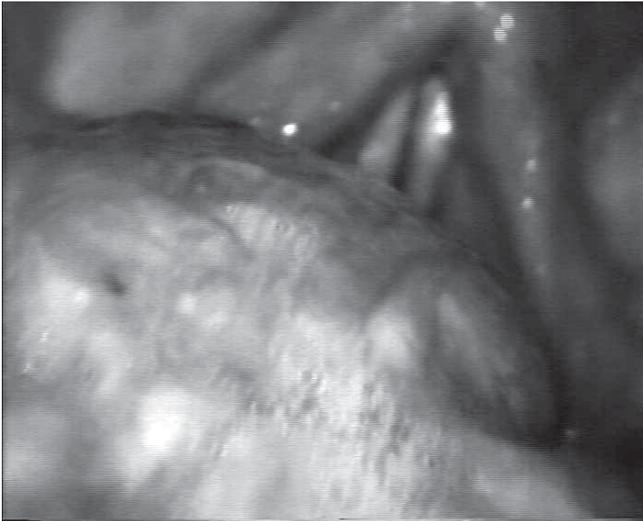


Fig. 1. Pre-operative laryngoscopy.

lar or tubular pattern of growth, at times forming papillary structures growing in the cystic cavities. Neoplastic cells showed immuno-histochemical positivity for S100 protein, actin and cytokeratin. Morphological features and the immuno-histochemical profile were coherent with an epithe-

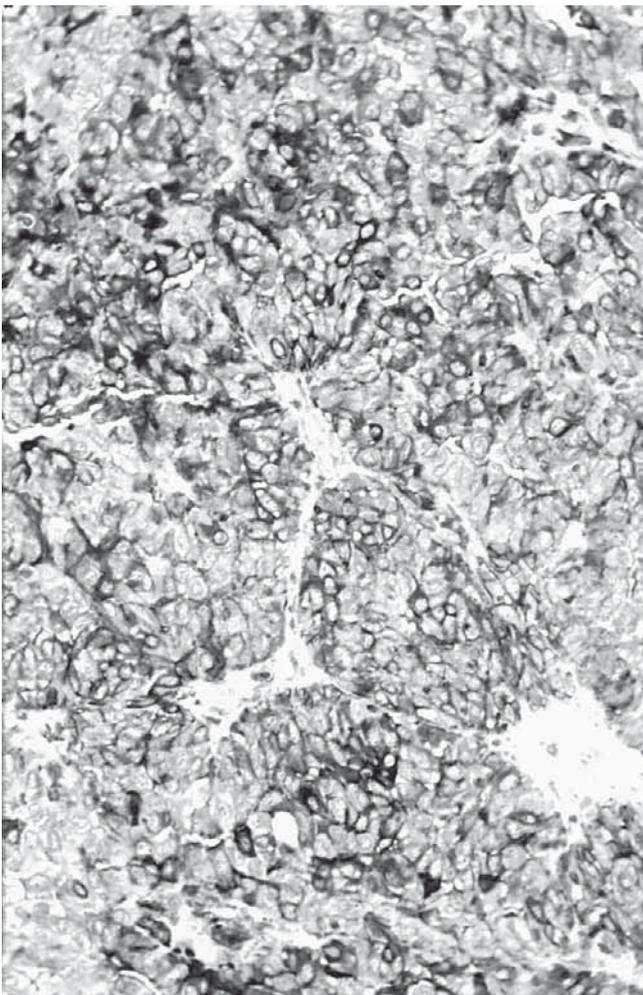


Fig. 2. Histological findings.

lial-myoepithelial carcinoma, derived from the salivary glands (Figs. 2, 3).

Magnetic resonance (MR) demonstrated an irregular aspect of the inferior portion of the tongue base, more evident to the right, with structural alterations of the bundles of the homolateral genioglossus and geniohyoid muscles and reduced width of the pre-epiglottic space. In addition, the epiglottis appeared poorly dissociable and there was no evidence of lateral neck lymphadenopathy.

On the basis of this diagnosis, the patient decided to consult some Italian and foreign centres specialised in the treatment of head and neck tumours. The therapeutic options differed considerably:

- a) broadly demolitive surgery including partial glossectomy and total hypopharyngolaryngectomy. As a consequence, it would be necessary to perform a definitive tracheotomy and a free-flap of reconstruction to allow the ingestion of food;
- b) partial glossopharyngolaryngectomy surgery with temporary tracheotomy for respiratory assistance;
- c) a cycle of radiotherapy over the tumour region, three days a week for one month, for a total of 60 Gy.

In June 2000, the patient, confused by the previous suggestions, came to our attention, at the Operative Unit of E.N.T. of the "S. Maria della Pietà" Hospital of Nola. On the

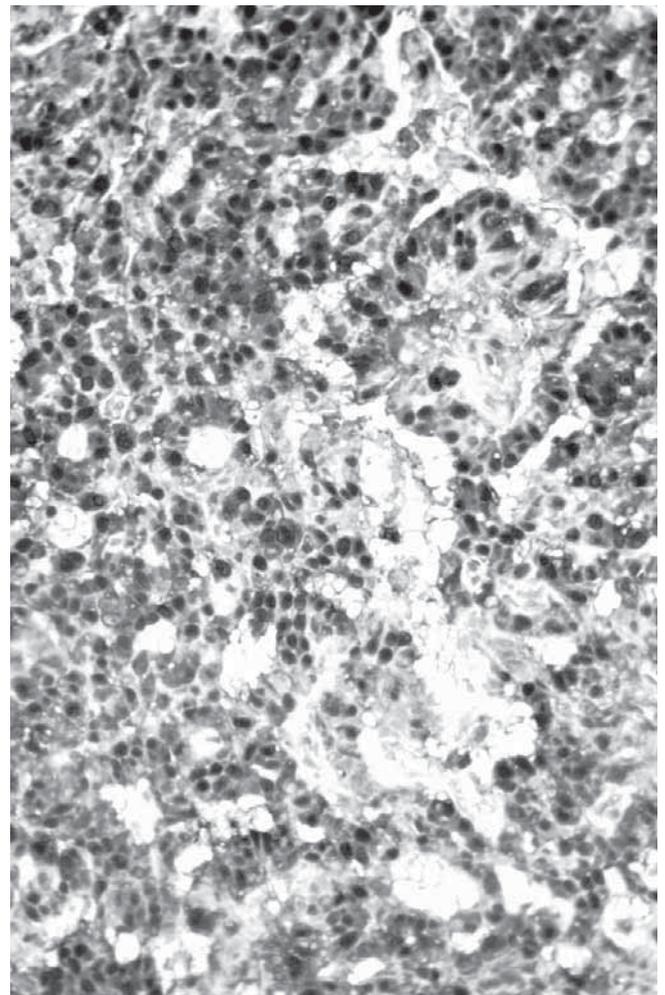


Fig. 3. Histological findings.

basis of the low-grade malignancy, confirmed at histological examination, and absence of recurrence, conservative treatment was decided in microlaryngoscopy exploiting the CO₂ Laser for the removal of the neoplastic mass. The patient decided to undergo this type of surgery despite the risk of local recurrence of the tumour about which we had duly informed her.

Upon hospitalisation, the patient had a slight mitro-tricuspidal insufficiency with normal pulmonary pressure. The anamnesis did not reveal cases of tumour; personal anamnesis was not relevant as far as concerns use of alcohol and tobacco; serology was positive for HCV.

Clinical examination, carried out upon direct laryngoscopy, the histological examination and the MR performed in our hospital substantially overlapped those previously performed.

Therefore, the patient was submitted to surgery with wide partial glossectomy extended to the pharyngoepiglottic folds and epiglottidectomy employing CO₂ Laser, followed by direct reconstruction of the tongue. The last step was introduction of a nasogastric tube to allow feeding through the natural approach in the immediate post-operative period. After 7 days the patient was able to swallow, without prob-

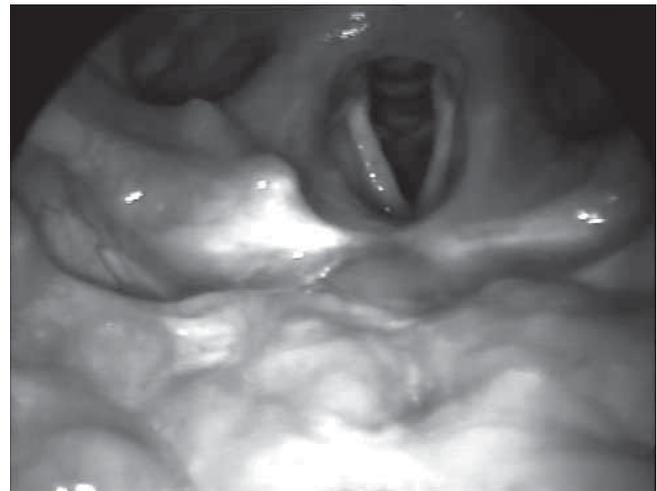


Fig. 6. Post-operative laryngoscopy.

lems, allowing us to remove the nasogastric tube. One week later, the patient was discharged from the hospital, on the way to recovery and able to eat by herself. Six months later, a control MR (Fig. 4) revealed normal morphology and signal of the muscular structures of the tongue and of the submandibular region; the aspect of the muscular-adipose plans of the para-oro-pharyngeal spaces appeared regular with the absence of lymphadenopathy.

Monthly controls were performed, through direct laryngoscopy and MR (Figs. 5, 6) that, to date, have confirmed perfect recovery of the surgical region and absence of recurrence after 30 months' follow-up.

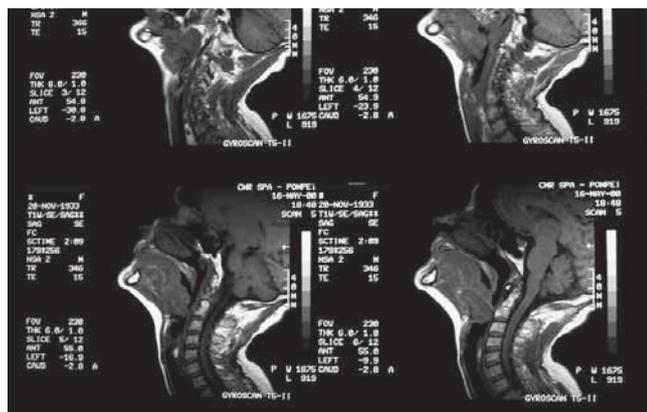


Fig. 4. 6 months post-operative MR.

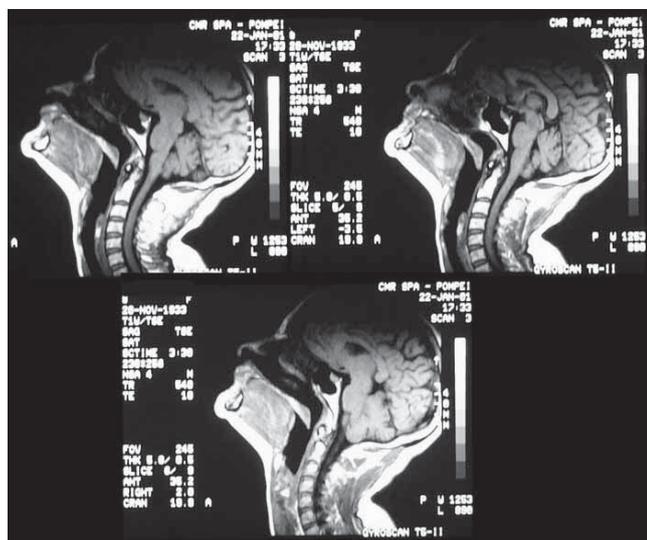


Fig. 5. Post-operative MR.

Discussion

Epithelial-myoeplithelial carcinoma of the salivary glands is a rare neoplasm, described for the first time by Donath et al. in 1972⁷. This tumour, reported by many authors with several different terminologies, had often been considered benign until consistent studies carried out with an appropriate follow-up recognized its metastatic capacity. This tumour has reached a definitive pathological entity in the international histological classification WHO of tumours in 1991⁸. However, it is still difficult to identify the histological differential diagnosis with other tumours of the salivary glands. This is due to the extreme variability of the relationship between myoeplithelial and ductal cells of the different epithelial-myoeplithelial carcinomas¹.

In spite of the recent appearance of this tumour, it was possible to identify its incidence in 1% of all tumours of the salivary glands, with a peak in the seventh decade of life and a slight (around 60%) predominance for the female sex. This neoplasm occurs primarily in the greater salivary glands (85-90%) and, particularly, the parotid. To date approximately 125 cases of epithelial-myoeplithelial carcinomas have been described, the greater part of which located in the salivary glands with the exception of some rare cases found in the nasal cavity⁴, paranasal sinuses⁵, lacrimal glands⁶ or bronchus and lung^{9,10}. However, location in the minor salivary glands of the base of the tongue has never been described in the literature.

Clearly, the prognostic and therapeutic aspects of this tumour represent the the most complex aspect of the case reported here.

Reports in the literature ¹¹ generally agree to consider epithelial-myoepithelial carcinoma as a low grade malignancy tumour, while the percentages of loco-regional recurrences and/or distant metastases are extremely variable. Brocheriou et al. ¹², in a review of the 71 cases diagnosed up to 1991, found 28% of local recurrences (20/71) and 8.5% of distance metastasis (6/71). On the contrary, Fonseca and Soares ², in a retrospective study of 22 cases, reported approximately 50% of local recurrences, in agreement with Batsakis ³, who considers this tumour has a low mortality but a strong loco-regional aggressiveness. In addition, these same Authors showed a metastasis rate of 35% with 40% of patients having died due to the neoplastic disease.

There are no clear data concerning the presence of indicative prognostic factors, even if Fonseca and Soares ² showed that nuclear atypias in more than 20% of the tumour cells could represent a morphological feature correlated to a negative prognosis. Finally, the treatment of choice, is predominantly surgical even if, obviously, determined by the structures involved by the tumour and by its extension. Adjuvant radiotherapy has been applied in only a few cases ¹³; on the contrary, there are no cases, in the literature, treated with a neoadjuvant radiotherapy.

The case reported here presented reassuring evidence, both histological and clinical, such as few atypias, rare mitotic activity and absence of lateral neck lymphadenopathy. On

the basis of these data and thanks to the CO₂ Laser, we have been able to programme and perform conservative endoscopic surgery that has probably allowed complete excision of the tumour thus avoiding the patient becoming an invalid after surgery. Undoubtedly, the association of a rare tumour in a location never previously found could have suggested to some surgeons the need to proceed with aggressive surgical techniques, with excision of a wide area to ensure total removal of the tumour. However, the validity of radiotherapy in the treatment of low grade malignancy tumour is very doubtful since this tumour will probably be of the low-responsive type.

In conclusion, the great disagreement regarding *quoad vitam* prognosis data – distant metastasis has been described 28 years after surgery ¹⁴ – suggests that 30 months' follow-up could be too brief a period to be sure of total recovery of the patient.

However, in our opinion, due to the uncertain evolution of this kind of tumour, a preferential approach should consist in the application of CO₂ Laser to perform conservative surgery while assuring radical excision of the tumour. In fact, recently, endoscopic surgery has gained more and more attention offering major benefits compared to traditional surgery, and, in this peculiar case, it might be strongly recommended since traditional surgery will lead the patient post-operatively to a poor quality of life.

Nevertheless, close and prolonged follow-up are mandatory in this therapeutic strategy in order to reveal, in an early stage and, consequently, treat, any local recurrence.

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Address for correspondence: Dr. E. Esposito, via Luca Giordano 26, 80127 Naples, Italy. E-mail: erik@unina.it