

Full Length Research Paper

## Salivary gland cystadenocarcinoma of the tongue: A case report and literature review

Mona Mlika<sup>1\*</sup>, Ines Chelly<sup>1</sup>, Heifa Azouz<sup>1</sup>, Houda Ouertani<sup>2</sup>, Ghazi Besbes<sup>2</sup>,  
Emna Mnif<sup>3</sup>, Slim Haouet<sup>1</sup>, Mohamed Moncef Zitouna<sup>1</sup> and Nidhameddine Kchir<sup>1</sup>

<sup>1</sup>Department of Pathology, La Rabta Hospital, Bab Saadoun, Tunis, Tunisia.

<sup>2</sup>Department of Otorhinolaryngology, La Rabta Hospital, Bab Saadoun, Tunis,

Tunisia. <sup>3</sup>Department of Radiology, La Rabta Hospital, Bab Saadoun, Tunis, Tunisia.

Accepted 2 September, 2013

**Cystadenocarcinoma is a rare salivary gland tumor which commonly arises in the ovary and the appendix. Only 7 cases have been reported in the tongue with 2 cases described as highly malignant. We report a case of cystadenocarcinoma of the base of the tongue in a 79-year-old man who presented with goiter and dysphagia. Physical examination showed a mass of the tongue. Treatment consisted in a partial glossectomy. The patient was free of recurrences after one year of follow up. Cystadenocarcinoma of the tongue is a very rare tumor on which diagnosis is based on histological findings. The treatment is based on surgical excision. These tumours are reported to have a good prognosis but some cases have been described reported to recur even after many years of follow up.**

**Key words:** Cystadenocarcinoma, base of the tongue.

### INTRODUCTION

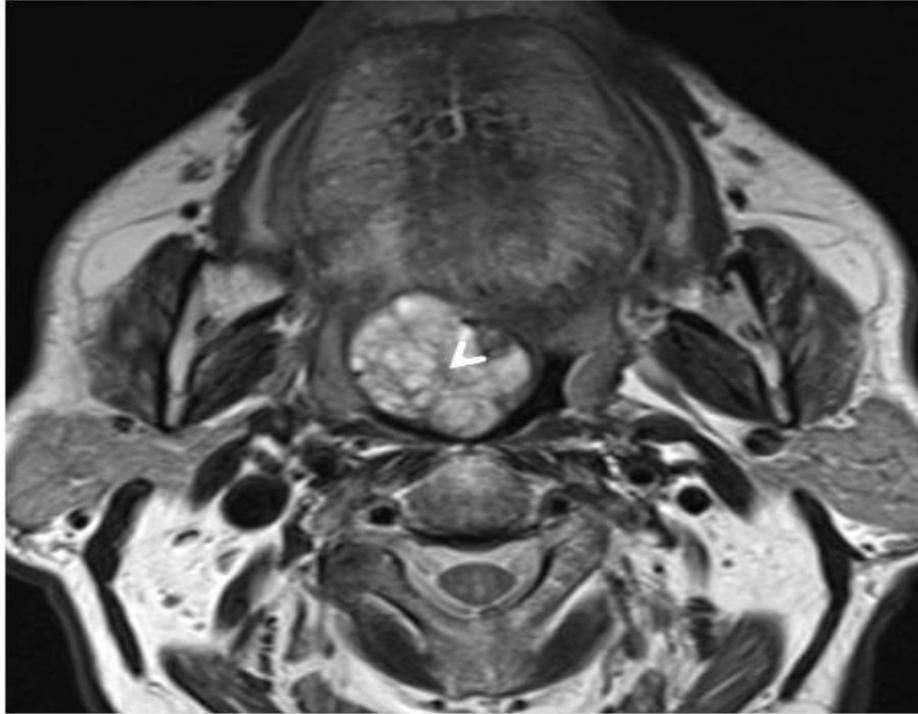
The most commonly diagnosed tumor of the tongue is the squamous cell carcinoma. The malignant salivary gland tumours of the tongue are very uncommon and are dominated by the low grade mucoepidermoid carcinoma followed by the adenoid cystic carcinoma. Papillary cystadenocarcinoma is an extremely rare malignant neoplasm first classified as a distinctive neoplasm in 1990 by the World Health Organization (Eveson et al., 2005). The authors describe a new case of cystadenocarcinoma of the tongue which is to the best of our knowledge, the eightieth reported case in the English literature. A 79-year-old male presented with goiter and dysphagia. He had history of hyperuricemia on medical treatment and history of appendectomy and cholecystectomy. Intraoral examination showed a submucosal nodule in the base of the tongue which was firm in consistency and adherent to the tongue musculature. There was no neck lymphadenopathy. Thyroid examination showed a retrosternal goiter. Cardio-vascular examination, pulmonary examination and abdominal

examination were normal. The general state of the patient was good (Karnofsky index: 90%). The CT-scan and the Magnetic resonance imaging (MRI) of the oral cavity showed a 3 cm lesion at the base of the tongue and demonstrated a loculated, multicystic appearance on T2 weighted sequences (Figure 1).

These findings were suggestive for a pleomorphic salivary gland adenoma. Chest x Ray and abdomino-pelvic CT-scan were normal. An incisional biopsy was performed and histology showed a tumour with cystic cavities infiltrating the tongue musculature. Some cysts showed papillary projections and the cell lining was columnar with uniform nuclei. A special stain with the periodic acid-Schiff showed the mucous secretion by the tumor cells (Figures 2A, B and C). An immunohistochemical study was performed and showed the expression of high molecular weight cytokeratin, S100 protein and negative for smooth muscle actin antigen by tumour cells (Figure 2D) with these histological and immunohistochemical findings we had an impression of acinic cell carcinoma with a papillary cystic growth pattern but there was no acinar differentiation.

The diagnosis of mucoepidermoid carcinoma was excluded because there were no typical histological characteristics and finally the diagnosis of salivary duct

\*Corresponding author. Email: [mlika.zorgati.mona@hotmail.com](mailto:mlika.zorgati.mona@hotmail.com).



**Figure 1.** MRI of the oral cavity showing a well circumscribed mass of the base of the tongue with small cystic cavities (arrow).

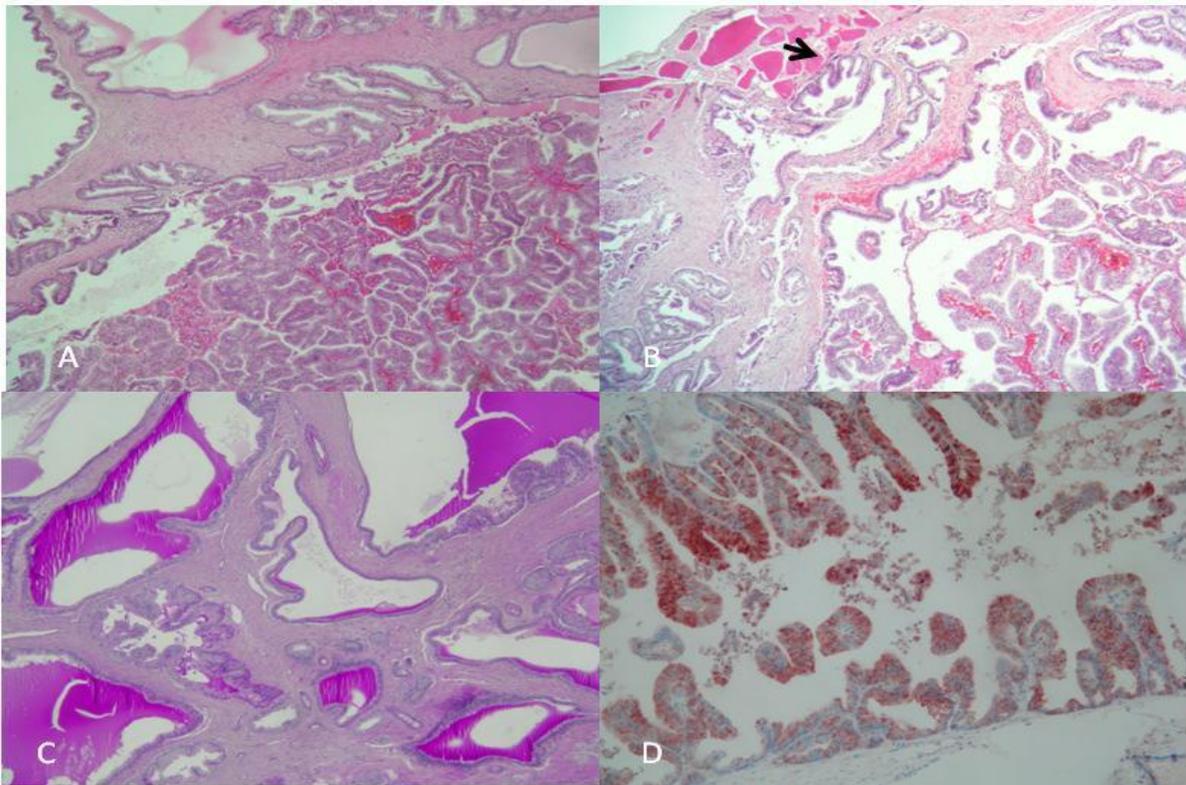
carcinoma was The diagnosis of polymerphous low grade adenocarcinoma was not retained because there was no particular architectural pattern. The cystadenoma was excluded because of the infiltration of the adjacent tissue. A metastatic papillary carcinoma from the thyroid gland was excluded because the histological examination of the thyroid specimen demonstrated a colloid goiter. Metastasis of an adenocarcinoma from the ovary, the intrahepatic bile duct, the pancreas or the gastrointestinal tract was ruled out because of the normal radiological findings. Based upon the above findings, low-grade papillary cystadenocarcinoma of the base of the tongue was diagnosed. This tumor was evaluated to be in an early stage (Stage 1) according to the clinical and radiological findings. The tumour mass was totally excised and the patient did not present recurrences within one year of follow up.

## DISCUSSION

Cystadenocarcinoma is a rare salivary gland tumour which commonly arises in the major salivary glands, mainly the parotid gland Metgud et al. (2007). Only 7 cases have been reported in the tongue with 2 cases described as highly malignant (Table 1). The 2 largest series about salivary gland tumours are those of Foss and coworkers with 57 cases and Goldblatt and

colleagues with 55 cases (Foss et al., 1996; Goldblatt et al., 1987). Only one case of cystadenocarcinoma of the tongue has been reported in each study. Some authors reported that these tumours may arise from excretory duct reserve cells (Metgud et al., 2007). The mean age of the patients was 74 years (average, 70 – 80 years). Most of the patients described were men who presented with a tongue mass in all cases. Radiologic examination was based upon CT scan and MRI findings. In spite of their necessity for confirming the presence of a mass, exploring its nature, showing the margins and revealing a lymph node enlargement, most of the time they are non specific (Grunstein et al., 2006).

In our case, there was no apparent gross invasion of the surrounding tissue making the radiologist evoke the diagnosis of pleomorphic adenoma. Positive diagnosis is based on pathologic findings. The definite means of diagnosis is the biopsy which may be challenging when it is superficial and cannot show the infiltration of the adjacent tissues on surgical specimen. Histological examination shows generally a well circumscribed but non encapsulated mass with numerous cysts partially filled with mucin and small solid islands or duct-like structures with varying degree of papillary proliferation (Nakagawa et al., 2002). Two variants have been reported by some authors: the low grade and the high grade. Low grade tumours are defined by uniform nuclei and rare mitoses (Grunstein et al., 2006). The tumour described in the



**Figure 2.** (A) Tumour proliferation formed by cystic cavities filled with papillary projections (HE  $\times 200$ ). (B) Infiltration of the tongue musculature by the tumour (arrow) (HE  $\times 200$ ), (C) Special coloration with the periodic Schiff-acid showing the mucous secretion, (D) Tumour expression of the cytokeratin by the tumour cells (HE  $\times 200$ ).

**Table 1.** Cases of cystadenocarcinoma of the tongue published in the English literature.

Authors	Year of publication	No. of cases	Sex	Symptoms	Treatment	Follow-up
Goldblatt et al.	1987	1	Male	Mass of the tongue	Surgical resection	-
Foss et al.	1996	1	Male	Mass of the tongue	Surgical excision	GGH (59 months)
Pollett et al.	1997	1	Male	Mass of the tongue	Partial glossectomy and neck dissection	Recurrences (6 months)
Nakagawa et al.	2002	1	Male	Mass of the tongue	Partial glossectomy	-
Spinato et al.	2002	1	Male	Mass of the tongue	Surgery with radiation therapy	-
Grunstein et al.	2006	1	Male	Mass of the tongue	Surgery	-
Metgud et al.	2007	1	Female	Mass of the tongue	Surgical excision and neck dissection	GGH (1 year)
The present case	2010	1	Male	Mass of the tongue	Surgical excision	GGH (1 year)

Nbre: Number, GGH: Good general health.

observation seems to share these characteristics. High grade tumours are characterized by nuclear pseudo stratification, high nuclear cytoplasmic ratio, numerous mitotic figures and the presence of areas of necrosis (Spinato et al., 2002).

The most striking feature to make the diagnosis is the infiltration to the adjacent tissues. Immunohistochemical study is not mandatory to make the diagnosis but some authors reported a positivity of the tumour cells with the cyokeratin, smooth muscle actin and S100 protein antigens (Pollett et al., 1997). Differential diagnoses include cystadenoma but this tumour is characterized by the absence of the infiltration of the adjacent tissue, a low grade mucoepidermoid carcinoma which may be cystic but is characterized by the presence of squamoid cells with mucus production and cells of intermediate type (Eveson, 2005). A papillary variant of acinar cell carcinoma may also be suspected but the most striking feature is the presence of acinar differentiation which is absent in cystadenocarcinoma (Grunstein et al., 2006). In our case, all these diagnoses were systematically excluded. The treatment is based on surgical excision associated to neck dissection and postoperative radiation therapy for neck metastases (Metgud et al., 2007).

## CONCLUSION

Cystadenocarcinoma is a very rare tumor of the salivary glands. Their occurrence in the tongue from the minor salivary glands is exceptional. These tumours have been reported to be of good prognosis but a close follow up is necessary because of their propensity for local recurrence and metastases even after many years.

## REFERENCES

- Eveson JW, Auclair P, Gnepp DR, El-Naggar AK (2005). Tumours of the salivary glands. In: WHO Pathology and genetics of head and neck tumours. Lyon, 209-236.
- Foss RD, Ellis GI, Auclair PL (1996). Salivary gland cystadenocarcinoma. A clinicopathologic study of 57 cases. *Am. J. Surg. Pathol.*, 20: 1440-1447.
- Goldblatt LI, Ellis GL (1987). Salivary gland tumours of the tongue. Analysis of 55 new cases and review of the literature. *Cancer.*, 60: 74-81.
- Grunstein E, Kacker A (2006). Salivary gland cystadenocarcinoma of the mobile tongue, low-grade papillary adenocarcinoma variant: a case report. *Ear Nose Throat J.*, 85: 829-830.
- Metgud R, Kalburge J, Dongre S, Karle R (2007). Papillary cystadenocarcinoma of the tongue. *JOMFP.*, 11: 63-65.
- Nakagawa T, Hattori K, Iwata N, Tsujimura T (2002). Papillary cystadenocarcinoma arising from minor salivary glands in the anterior portion of the tongue: A case report. *Auris Nasus Larynx*, 29: 87-90.
- Pollett A, Perez-Ordenez B, Jordan RC, Davidson MJ (1997). High-grade papillary cystadenocarcinoma of the tongue. *Histopathol.*, 31: 185-188.
- Spinato R, Abramo A, Manconi R (2002). Highly malignant papillary cystadenocarcinoma of the tongue: a case report. *Acta Otorhinolaryngol. Italy*, 22: 86-89.