MYOEPITHELIAL CARCINOMA OF THE SUBMANDIBULAR GLAND: A RARE CASE WITH LUNG METASTASIS

CARCINOME MYOÉPITHÉLIAL DE LA GLANDE SUBMANDIBULAIRE: UN CAS RARE AVEC MÉTASTASE PULMONAIRE

H. Hablani[1] *, S. Briki[1], W. Elleuch[1] , F. boudawara[1], J. Bennoun[1], A. Hentati[2], F. Karray[1] , M. Abdelmoula[1]

[(1)]: Oral and maxillofacial Surgery department, Habib Bourguiba Hospital university, Sfax, Tunisia

[(2)]: Thoracic Surgery department, Habib Bourguiba Hospital university, Sfax, Tunisia

ABSTRACT

Introduction: Myoepithelial carcinoma of the submandibular gland is uncommon. Its clinical presentation is nonspecific, making diagnosis and treatment challenging.

Case report: We report a case of a 52-year-old male presenting with a painless swelling in the floor of the mouth. Clinical and imaging studies initially suggested cystic lymphangioma. Further investigations concluded to myoepithelial carcinoma with lung metastasis. Large tumor resection via a transmandibular lateral pharyngectomy approach was performed, along with a right upper lung lobectomy. Despite aggressive surgery and adjuvant chemoradiotherapy, local recurrence occurred.

conclusion: This case highlights myoepithelial carcinoma's aggressive nature. Vigilant surveillance is imperative, though the risk of recurrence remains high.

Keywords: Myoepithelial Carcinoma, Submandibular Salivary Gland, Lung Metastasis, Surgery, Local recurrence

RÉSUMÉ

Introduction: Le carcinome myoépithélial de la glande submandibulaire est rare. Sa présentation clinique est non spécifique, ce qui rend le diagnostic et le traitement difficiles.

Cas clinique: Nous rapportons le cas d›un homme de 52 ans présentant une tuméfaction indolore au niveau du plancher buccal évoquant le diagnostic d'un lymphangiome kystique. Les explorations ont conclu à un carcinome myoépithélial avec métastase pulmonaire. Une résection tumorale large par la voie bucco-pharyngectomie trans-mandibulaire a été réalisée, ainsi qu›une lobectomie pulmonaire supérieure droite. Malgré une chirurgie agressive et une chimio-radiotherapie adjuvante, une récidive locale est survenue.

Conclusion: le carcinome myoépithélial submandibulaire est une tumeur agressive. Une surveillance vigilante est impérative, vu le risque de récidive élevé.

Mots-clés: Carcinome myoépithélial, Glande submandibulaire, Métastase pulmonaire, Chirurgie, Récidive locale.

INTRODUCTION:

Myoepithelial carcinoma (MC) is a rare salivary gland malignancy that was first described in 1975 by Stromeyer et al. [1] and has been included in the World Health Organization (WHO) classification of salivary gland tumors since 1991 [2]. While most cases affect the parotid gland, submandibular involvement is uncommon. Diagnosis relies on histopathology due to a nonspecific clinical presentation. MC demonstrates aggressive behavior with frequent local recurrence and potential metastasis to sites such as the lungs, bone, and brain [3]. Treatment primarily involves wide surgical resection, though optimal management strategies remain undefined.

This report presents a rare case of submandibular MC with synchronous lung metastasis and discusses therapeutic approaches and prognostic factors based on a literature review.

CASE REPORT:

A 52-year-old male presented with a two-year history of a painless, gradually enlarging floor-of-mouth swelling. Extraoral examination showed a slight swelling in the right submandibular region, while intraoral assessment revealed a firm, well-defined 40 x 20 mm mass that displaced the tongue and was covered by normal mucosa. No lymph nodes were palpable in the cervical area (Figure 1).

corresponding author:Hanen Hablani
E-mail: hablanihanene@yahoo.fr

Figure 1: A 52-year-old patient presented with a volume mass of the floor of the mouth, pulling the tongue.

Computed tomography (CT) of the submandibular region revealed a cystic, multiloculated collection in the submandibular gland with well-defined margins, measuring 40 mm by 22 mm in the axial plane. The walls enhanced after the injection of a contrast medium, and there was no associated bone lysis or cervical lymphadenopathy. The diagnosis of cystic lymphangioma of submandibular gland was suspected (Figure 2).

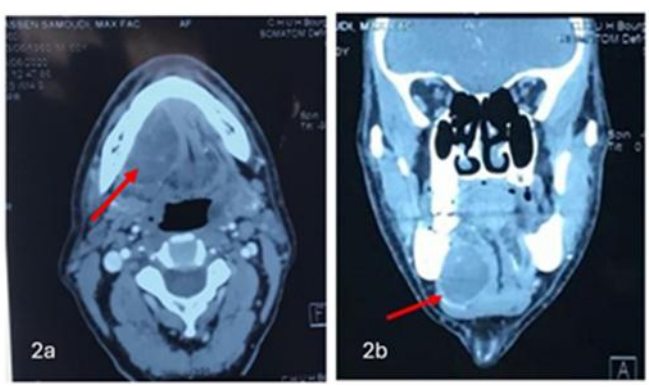

Figure 2: Head and neck Computed Tomography in axial (2a) and coronal section (2b) revealed a cystic, multiloculated collection in the submandibular gland with well-defined margins, measuring 40 mm by 22 mm in the axial plane, without adjacent bone lysis.

During the pre-surgery investigation, an irregular opacity in the right lobe of the lung was discovered in a chest X-ray. A CT scan was subsequently performed, revealing a suspicious mass in the right upper lobe (Figure 3).

Figure 3: A chest X-ray (3a) and axial CT scan (3b) revealed a solitary suspicious lesion in the right upper lobe.

A biopsy of the lung tumor was indicated and performed. It showed a metastatic MC, originating from the submandibular gland. Immunohistochemical staining revealed positive expression of keratin, vimentin, and EMA.

A bone scan was negative for further metastases, resulting in a final staging of T3 N0 M1.

According to the head and neck oncology committee of our institution, treatment is based on surgery. The trans mandibular lateral pharyngectomy approach was chosen to better expose the tumor. It allows good control over the total resection. The mandibular inner cortex was resected with the tumor. Intraoperative frozen section analysis confirmed negative margins at all resection limits. An ipsilateral selective neck dissection was also performed. Reconstruction of the mucosal defect of the floor of the mouth was achieved using a myo-mucosal buccinator flap (fig 4). A right upper lung lobectomy was performed simultaneously.

Figure 4: Operative view of tumor resection and defect reconstruction. (4a): Good exposure of the tumor through the transmandibular lateral pharyngectomy approach. (4b): Reconstruction of the mucosal defect by a myo-mucosal buccinator flap.

Definitive histopathological and immunohistochemical examinations confirmed the diagnosis of MC of the submandibular gland. The Ki-67 proliferation index was 25%, with necrosis and perineural invasion, and no evidence of vascular emboli. All resection margins were negative, and adjuvant chemoradiotherapy was indicated.

The patient died from a local tumor recurrence 22 months later.

DISCUSSION

Myoepithelial carcinoma (MC) is a rare malignancy, representing less than 1% of all salivary gland cancers. It most frequently occurs in the parotid gland, while submandibular gland involvement is uncommon [1,3,4]. According to the WHO, these tumors can arise de novo or from pre-existing conditions (pleomorphic adenoma or myoepithelioma) [5,6,7,8].

Patients are typically diagnosed between their third and fifth decades of life [3,4], with no significant sex predilection. Clinically, most patients present with a non-specific, slow-growing, painless mass [1,6]. These non-specific clinical signs often lead to misdiagnosis. In the present case, the patient presented with a painless, well limited mass in the floor of the mouth, which was clinically and radiologically misdiagnosed as a cystic lymphangioma.

A definitive diagnosis relies on immunohistochemistry. It tests positive for specific markers like vimentin, cytokeratin (CK), epithelial membrane antigen (EMA), CD10, smooth muscle actin (SMA), S100 protein, and p63 [2,4].with tumors typically showing immunopositivity for markers such as vimentin, cytokeratin (CK), epithelial membrane antigen (EMA), CD10, smooth muscle actin (SMA), S100 protein, and p63 [2,4].

Surgery is the main treatment for myoepithelial carcinoma of salivary glands, resection must be large to get R0 margin. Given the low rate of lymph node involvement, routine neck dissection is not indicated unless nodal disease is confirmed [5,7].

The role of adjuvant therapy remains unclear. The authors indicate that this additional radiation did not improve cancer control or survival rates. Reports by Nieder et al. [9] have shown that radiation therapy following incomplete surgery also did not help in controlling cancer or enhancing survival. In our case, adjuvant chemoradiotherapy was indicated for a large tumor size, presence of metastasis in the lung, and perineural invasion.

Recurrences and metastasis can occur anywhere from several months to many years after the initial resection. In our case, the patient was first presented at the metastatic stage. This is due to the slow progression of the tumor and its indolent characteristic, which led the patient to consult in the advanced stage of the disease. See et al [10] described in his study the risk factors and prognosis for MC of the major salivary gland, he concludes that there's no difference in clinical outcome of MC "de novo" versus those that arise in pleomorphic adenomas and/or myoepitheliomas. Meanwhile, MC, carries a relatively high risk of local-regional recurrence and distant metastasis. He also identified that age under 60 is a predictive factor for distant metastasis. Other histological features like nuclear atypia, necrosis, and an elevated mitotic count can contribute to recurrence. From the few cases of the literature, authors [8,10] reported that positive margins, angiolymphatic invasion, necrosis, and myoepithelial immunohistochemistry staining positivity for S-100 and Ki-67 are predictors of decreased disease-free survival. Additionally, a tumor size greater than 4 cm and high-grade histology were associated with increased mortality in 6.5% of patients.

CONCLUSION

Myoepithelial carcinoma (MC) of the submandibular is an aggressive malignancy with high risks of recurrence and metastasis. This case underscores the diagnostic challenges, especially with synchronous lung metastasis. Accurate diagnosis requires comprehensive histopathological and immunohistochemical evaluation, while surgical resection with clear margins is the main treatment.

Conflict of interest:

There is no conflict of interest.

Ethical approval: The patient provided written informed consent for publication.

Funding: The authors declare that no specific funding was received for this work.

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