

Macroglossia caused by adenoid cystic carcinoma. Case report

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Abstract

A 33-year-old woman sought medical care at our institution for macroglossia. She had been treated before with tracheostomy and gastrostomy due to an adenoid cystic carcinoma (ACC) of the mobile tongue, in clinical stage IVC. She was subjected to total glossectomy with larynx preservation, modified radical dissection of the right neck and of the left supraomohyoid muscle, as well as reconstruction using a free flap from the rectus abdominus. The histological report described a 15x11 cm solid type ACC of the mobile tongue (grade III), with infiltration to adjacent soft tissues, vascular and lymphatic vessels, as well as two metastatic lymph nodes in the right neck dissection. One month after surgery, bone and bilateral pulmonary tumor activity became evident. In advanced-stage tumors, treatment must be individualized, and when accompanied by severe deterioration of the quality of life, surgery is recommended independently from the extension, provided that a good reconstruction is accomplished.

Key words: Adenoid cystic carcinoma, tongue, microvascularized free flaps, macroglossia.

Introduction

Adenoid cystic carcinoma (ACC) is a rare malignant tumor, representing about 1 to 2% of all head and neck neoplasms and from 10 to 15% of all salivary gland neoplasms (1,2). The most common site for minor salivary gland neoplasms is the palate, followed by the base of the tongue, with a presentation rate of 8.8%; in contrast, the mobile tongue comprises only 2.9% of the cases (3,4). Regional metastases occur in less than 3%; however, distant metastases are more frequent and involve mainly the lungs and bones, followed in descending order by the brain, liver, and omentum (5).

Histologically the pattern is classified as solid, tubular or cribriform; perineural invasion is common and has been reported in almost 50% of the cases; less frequently, necrosis and vascular invasion have also been reported (5). The solid pattern is the one with the worst prognosis (6). The mainstay of treatment is surgical resection, and some authors suggest

the use of adjuvant radiotherapy (7). We present a case of macroglossia due to ACC with advanced-stage disease.

Case report

A 33 year-old female patient started her illness 10 months before admittance to our institution with a progressive tumoral growth of the mobile tongue that protruded from the oral cavity. It produced mandibular pain due to inability to attain occlusion. Four months before she was submitted in another hospital to emergency tracheostomy due to airway obstruction, gastrostomy and neck biopsy, with a report of adenoid cystic carcinoma (Figures 1a and b). The staging according to AJCC (8) corresponds to T4a N2c M1, clinical stage IVC. One day after her admittance to our institution, she was submitted to surgery, consisting of total glossectomy with preservation of the larynx, modified radical dissection of the right neck and the left supraomohyoid muscle, as well as reconstruction with a

free rectus abdominus flap, choosing the contralateral side of the gastrostomy (Figure 2). The patient evolved satisfactorily and was discharged from the hospital on the 10th postoperative day. The histopathologic report described a 15x11 cm solid type ACC of the mobile tongue (grade III), with infiltration to soft tissues and vascular and lymphatic structures, as well as two metastatic ganglia in the right neck dissection.



Fig. 1. a and b. Frontal and lateral views of the patient with macroglossia due to ACC.



Fig. 2. Right donor site of the rectus abdominus in the abdomen.

One month after surgery she reported intense left subclavicular pain which increased toward the left sternoclavicular junction, where a small mobile and painful nodule could be palpated; she also had persistent cough and nausea. The diagnosis of bone and bilateral pulmonary tumoral activity, which had been diagnosed earlier, was then confirmed. She was subjected to thoracic computed tomographic scan (CT) and bone gammagraphy, and was programmed to receive palliative treatment. However this could not be accomplished because the patient was lost for treatment and follow-up at 2 months and 10 days after surgery.

Discussion

ACC is an infrequent lesion of the head and neck, which accounts for 1 to 2% of all neoplasms in this anatomic region. Tongue involvement by ACC has been reported in 3 to 11% of all the cases that occur in the head and neck, and 11.9% of the cases seen at our institution. The base of the tongue is the second most frequently affected intraoral site by minor salivary gland neoplasms, with approximately 20% of the cases, only outnumbered by the soft palate (4), and among these, ACC represents 30%. Hence, our case represents even a smaller percentage of these tumors, since its presentation in the mobile tongue is extremely rare (less than 3% (3,4)). The clinical data presented by patients are relatively non-specific, as it tends to occur as a painless or asymptomatic submucosal growth, which hinders an early diagnosis. According to previous reports, time elapsed from the first clinical manifestation until the presence of symptomatology ranges from 2.5 to 7 years (6), which contrast with our case, as it was of only 10 months, suggesting an extremely rapid growth.

The treatment of choice is surgery, although some authors recommend radiotherapy as adjuvant in patients with advanced T stage and/or positive surgical margins (7), or as single treatment in cases of surgical irresectability. Extension of surgery depends mainly on the T stage at diagnosis, that is, hemiglossectomy can be performed or up to total glossectomy with/without total laryngectomy and reconstruction with free microvascularized flaps. The purpose of placing a microvascular flap is to preserve the contact toward the palate and to mobilize food toward the oropharynx with a lower degree of aspiration. The use of an antebrachial, pectoralis major, wide dorsal, and rectus abdominus rectum flaps represents an option. Some disadvantages of pedunculated flaps, such as those from the pectoralis major are: dependence on its pedicle that hinders its correct mobility, the risk of distal marginal necrosis of the skin, and a lack of adequate volume(9-10). Microvascularized flaps, which in our experience provide an adequate volume, are those from the rectus abdominus and the wide dorsal, in contrast to the antebrachial flap or the pedunculated flaps from the pectoralis major (11) (Figure 3).



Fig. 3. Final reconstruction after total glossectomy with larynx preservation. One can see a perfect palatoglossal contact achieved through a bulky free flap.

It has been reported that 75% of the cases of ACC located at the base of the tongue are in stages T3 and T4 (12) at the time of seeking medical care; however, those presenting in the mobile tongue, are more frequently seen in earlier stages. In our case, the patient sought medical care at a late stage (IVC), most probably because she came from a low socioeconomic level and because of the absence of early neurological symptoms, which delayed her need for seeking medical assistance.

This case merits considerations and questioning from the point of view of the treatment that should be provided in such an extraordinary example of ACC with an advanced disease, where the clinical dilemma is always survival and function when performing such an extensive surgical procedure with an equally extensive reconstruction, exerting a profound impact on speaking and swallowing despite being able to preserve the larynx. However, in this case, the patient had already required a gastrostomy and a tracheostomy before her admittance to our institution, with ankylosis of the temporomandibular joint secondary to the protrusion of the tumor, aside from the social confinement to which this patient was submitted due to her physical appearance. It is well known that patients with a lingual ACC are unlikely to be cured, and even more a patient like ours already suffering of metastatic pulmonary disease. However, as these patients can survive for a long time despite the metastatic disease, it was decided to provide surgical treatment aimed primordially at improving her quality of life, independently from her lifetime.

Conclusion

ACC remains a rare entity of the oral cavity, more so in the mobile tongue; however, treatment of advanced-stage tumors must be individualized due to the current lack of information because of the scarce number of similar cases. Surgery in advanced stages with a severe impact on the quality of life must be recommended without consideration to the extension as long as a good reconstruction is performed.

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