Giant macroglossia in head and neck cancer survivor: The unsolved issue

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ABSTRACT

Introduction: Most laryngeal cancers are squamous cell carcinomas. These tumors are often diagnosed in males over the age of 50, heavy smokers and drinkers. Thanks to the recent advances in multidisciplinary treatment overall survival has increased and thus the number of long-term survivors. Macroglossia may be secondary to head and neck cancer treatments occurring due to a blockage of the efferent lymphatic vessels and it is incurable. It seems to be an indolent complication, however, it can cause serious feeding difficulties, bleeding or ulcerations and subsequently infections with a reduction in quality of life and even survival. Case Report: A 72-year-old man heavy smoker was diagnosed with undifferentiated laryngeal squamous cell carcinoma stage III (T2N2bM0). He underwent an oncological surgery followed by concurrent chemoradiation therapy. Thirteen months after the patient complained about a slowly progressive tongue swelling with eating and speaking difficulties and sialorrhea. An otolaryngological exploration did not show any lesion but a tongue thickening with a massive enlargement was noticed. He was treated with diuretics, steroids and cervical massages with a slight objective improvement. The patient’s general condition deteriorated quickly due to a respiratory infection causing his death. Conclusion: Macroglossia secondary to lymphedema in patients with head and neck cancer warrants an intensified search for new treatment approaches, especially since survival of these patients has improved markedly. Complications secondary to this condition can be very relevant and sometimes can cause the patient’s death.

Keywords: Macroglossia, Larynx cancer, Facial lymphedema, Neck radiotherapy, Neck dissection.

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INTRODUCTION

Approximately one percent of all neoplasias arises from the larynx and laryngeal carcinomas comprises 45% of the head and neck cancers [1]. The majority of laryngeal carcinomas are squamous cell carcinomas [1]. This kind of disease is most often found in males over the age of 50 and it is often associated with heavy tobacco and alcohol use [1]. The most common
treatment plan is designed by a multidisciplinary team composed by all medical specialities involved in this cancer therapy such as radiotherapist, medical oncologist, otolaryngologist and radiologist, but most of these patients finally will receive the three treatment modalities: radiotherapy, surgery and chemotherapy [2]. Thanks to the advances in these treatments the overall survival and thus the number of long-term survivors has increased. This fact implies that quality of life has emerged as a new issue to be taken into account in all these patients and the multidisciplinary treatments could also impair this quality of life.

Macroglossia, also known as an enlarged tongue, is a component of numerous syndromes (list of most frequent syndromes is given in table 1), many caused by inherited metabolic anomalies in which the increase in tongue size is a manifestation of visceromegaly related to lysosomal storage diseases, such as Hurler syndrome [3]. Other macroglossia-associated disorders include Beckwith Wiedemann syndrome, neurofibromatosis type 1, amyloidosis, hemangiomatosis associated with Sturge-Weber syndrome, and congenital lymphangioma [3]. But this sign may be also secondary to head and neck cancers treatments such as radiation therapy or surgery [4]. This occurs because a blockage of the efferent lymphatic vessels. Although it seems to be a banal complication, it can cause serious problems of because the associated feeding difficulties and bleeding or ulcerations with secondary infections, which not only could impair the quality of life but also could decrease the chances of survival [4].

We present here one of these cases in which giant macroglossia appeared 13 months after finishing the treatment for laryngeal carcinoma in the context of facial oedema.

Table 1: List of more frequent causes of macroglossia.

- Down syndrome
- Multiple myeloma
- Hypothyroidism
- Acromegaly
- Amyloidosis
- Lymphoma
- Angioedema
- Hemangioma
- Tongue carcinoma
- Neurofibromatosis
- Down’s syndrome
- Lymphangioma
- Beckwith’s syndrome
- Congenital micrognathia
- Beckwith-Weidmann syndrome
- Congenital hypothyroidism
- Hurler syndrome
- Glycogen storage disease

CASE REPORT

A 72-year-old man with a long history of heavy smoking was admitted to our institution. The patient complained of progressive hoarseness, periodic difficulty in swallowing solid and liquid food, loss of appetite and weight for the last six months. Two months before consulting the patient had noticed pain in his right side of neck without any radiation and he referred to touch a painful and tough nodule in this area.

Physical examination showed enlarged cervical lymph nodes on the right side of the neck and laryngoscopic examination revealed a growth in the right vocal cord, ventral area of the epiglottis and adjacent area of the pharyngeal wall, with an irregular surface of approximately 4.5 cm × 3 cm × 2.5 cm in size. A biopsy of the growth was taken and an histopathologic diagnosis of undifferentiated squamous cell carcinoma was established. A computerized tomography did not show distant metastases, so a diagnosis of stage III cancer of the larynx (T2N2bM0) was made.

After obtaining informed written consent the patient underwent laryngectomy and radical surgery of bilateral cervical lymphnodes and a permanent tracheostomy. Then he received concurrent adjuvant chemoradiation therapy and finally entered in our program of periodic follow-up.

Thirteen months after the end of treatment the patient was referred to the outpatient clinic because of a slowly progressive swelling of his tongue in the last two and a half months, causing difficulty with eating and speaking and sialorrhea secondary to inability to swallow the saliva.

A neck and facial nuclear magnetic resonance imaging and computed tomography (CT) did not show any lesion suggesting local or regional recurrence but with voluminous distante disease (lung, mediastinum and liver) without any alteration in his analyses including thyroid hormone or tumoral markers. An exploration by the otolaryngologist did not show any lesion, of the two ways (digestive and airway), however a tongue thickening (Figure 1) with massive enlargement interfering with eating, swallowing and speaking but not with breathing was seen. The patient also presented with facial oedema mainly located at his eyelids with a fibrotic neck with a touch reminding of a tree trunk.

The patient could just tolerate swallowing of liquids with the help of a straw. He was treated with diuretics (furosemide 40 mg per day for one week and then 20 mg/daily) and steroids (hydrocortisone 400 mg/daily for 4 days and then 200 mg/daily for one week, then the dosage was reduced in 50 mg every four days) and cervical massages with a moderate subjective and a slight objective improvement within the first 15 days. The patient could finally tolerate semi-solid and liquid diet. But after several days the
patient’s general condition which had never been good with an ECOG (Eastern Cooperative Oncology Group) of 2-3, rapidly deteriorated because of aspiration pneumonia associated with severe respiratory failure unresponsive to oxygen therapy and broad spectrum antibiotic therapy. The patient died within a few hours after this complication (three weeks after being admitted to hospital).

DISCUSSION

The occurrence of facial edema with secondary macroglossia is a serious problem of patients suffering from cancer and may have various causes [4]. In several cases it indicates the progression of the illness but in others it can be secondary to treatments received. Regardless of its origin it results in unpleasant physical changes and functional impairments which add to the already existing issues caused by the cancer disease and treatments. So far therapeutic interventions are limited. Current management consists of physical therapy and pharmacological interventions trying to reduce swelling, restore shape, and prevent inflammatory episodes. Meticulous skin and mucosa care is needed to avoid recurrent inflammatory episodes and drug therapy has included the use of diuretics, corticosteroids, and coumarin- or flavonoid-type compounds.

Our patient received diuretics, steroids and massages showing just a little benefit. Despite of all these therapies much remain to study in searching for new and more active treatments [5]. There are some published clinical case reports describing the successful use of selenium and sometimes sandostatin to treat facial edema of a patient with advanced head-neck cancer but larger studies are required. In our case and due to his short survival we could not offer him a treatment with selenium which it had been planned previously.

Coumarin and flavonoids are other treatments aimed at reduce swelling in all types of lymphedema, but their long-term use is problematic [5].

In this context one promising step in therapy seems to be the introduction of free-radical scavengers such as selenium. This product can enhance the benefits of physical therapy in radiation-induced lymphedemas and its very low toxicity profile and cost effectiveness are further arguments for its use in lymphedema treatment.

CONCLUSION

Regardless of etiology facial lymphedema with secondary macroglossia is essentially incurable. The unsatisfactory status of lymphedema treatment in patients with cancer warrants an intensified search for new treatment approaches, especially since survival of cancer patients has improved markedly.

Authors Contributions
Esther Uña Cidón – Substantial contributions to conception and design, Acquisition of all data, Drafting the article, Revising it critically for important intellectual content, Collecting the photo, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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