TRIGEMINAL NEURALGY OCCASIONED BY CYSTIC ADENOID CARCINOMA IN PALATE REGION: CLINICAL CASE REPORT

NEURALGIA TRIGÊMINAL OCASIONADA POR CARCINOMA ADENÓIDE CÍSTICO NO PALATO: RELATO DE CASO CLÍNICO

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ABSTRACT

Adenoid cystic carcinoma (ACC) is the most common histological type among salivary gland neoplasms. Its growth is slow and steady and can vary from asymptomatic for months or even for some years, to extremely painful, which often leads the patient to seek treatment early. The palate is the most frequent intraoral location of these salivary gland tumors, representing approximately 50% to 60% of cases[31]. Its clinical appearance has a rounded shape and a smooth surface in most cases, or ulcerated, affecting mainly the posterolateral region of the palate. Patient S.F.A., female, 26 years old, came to the Head and Neck Surgery service of the Hospital dos Fornecedores de Cana complaining of a nodule in the region of the soft palate and intense odynophagia. During the anamnesis, the patient reports having noticed the injury 8 years ago and being a cocaine user, ex-alcoholic and ex-smoker, without other comorbidities. On physical examination, fibroelastic and submucosal lesions were noted, affecting the posterior region of the soft palate and the posterior portion of the left hard palate. She underwent a partial left maxillectomy for resection of a minor salivary gland tumor, selective cervical lymphadenectomy on the left and adjuvant radiotherapy.

KEYWORDS: Adenoid cystic, adenoid cystic carcinoma, maxillectomy e salivary gland tumor.

1. INTRODUCTION

Adenoid cystic carcinoma (ACC) was first described by Billroth (1854). This author applied the term “cylindroma” when presuming to be a pathology originated from nasal mucosa glands. Billroth designated this expression when referring to a structural quality of the tumor stroma and not to a particular epithelial component. Over time, this term fell into disuse, being replaced by other authors who described it as epithelial cells in a cylindermatous shape. The name adenoid cystic carcinoma was established years later by James Ewing. ACC can also be found in the literature by the names of cylinderma, adenoid cystic adenocarcinoma, epithelioma and adenomyoma-epithelioma[1]. Until 1940, this tumor was considered a benign variant of the mixed tumor of salivary glands, and Dockerty & Mayo (1943) used the term “malignant” to describe such lesion and indicated that this neoplasm had the peculiarity of spreading through the neural pathway[2].

ACC is responsible for 10% of neoplasms in the oral and maxillofacial region, and about 22% of malignant tumors of the minor salivary glands[3]. It is a tumor that affects mostly minor salivary glands, especially in the palate region, while it is less common in larger salivary glands (parotid and submandibular). Its prevalence is about 20% to 25% and 12% to 17% in these regions, respectively. Also, it can affect lacrimal glands, nasopharynx, trachea, serous glands and occasionally secretory glands of the female genital tract[4].

Tumor growth is slow and occurs through extensive perineural growth. Although hematogenous dissemination, in turn, occurs frequently in this disease. The treatment of choice consists of total resection of the lesion, with the benefit of adjuvant therapies being controversial[5]. The cyst is encapsulated and well circumscribed. Histologically, it is composed of a fusion of myoepithelial cells and ductal cells, with a highly variable proportion of these elements, with three variations: cribriform, tubular and solid. The ACC has an arrangement of small cells with little cytoplasm, surrounded by cell strands that may be empty or

contain mucus and / or hyaline or mucous material[6]. In most studies, the solid-state has the worst prognosis, associated with an advanced stage and the development of distant metastases. Solid lesions with areas of necrosis are associated with a more fulminating course[7].

Because ACC has slow and variable growth, Foote & Frazell (1953), reported in their unparalleled article on "Tumors of the major salivary glands" that the time for patient admission to the hospital varies from a few months to 20 years, with an average of 5 years. Quattlebaum, Dockerty, and Mayo (1954) reported, from their research, that the average time varies from 6 months to 15 years, with an average of 7 years[8].

The presente study aims to report the case of a patient with trigeminal neuralgia caused by cystic adenoid carcinoma on the palate, taking into account the treatment and prognosis.

2. CASE REPORT

Patient, 26 years old, caucasian, attended the Head and Neck Surgery service at the Hospital dos Fornecedores de Cana complaining of severe pain in the left hemiface and posterior palate region with difficulty swallowing. In anamnesis, the patient denies being a smoker but confirms that she is an alcoholic and has been using cocaine for a long time. She reports having noticed the appearance of the lesion about 8 years ago and progressive growth associated with increased pain for about 5 months. On physical examination, a lesion of fibroelastic consistency and dimensions of 2.8 x 2.7 x 2.2 cm was noted in the soft palate region, partially affecting the nasopharynx (Figure 1). Its surface was smooth, reddish in color and not ulcerated. The patient underwent Magnetic Resonance Imaging of the Face and Skull for staging and aspiration puncture with a fine needle (PAAF) of cervical lymph node for anatomopathological examination. Her surgery was performed on an urgent basis after successive visits to the emergency department and opioid abuse due to difficult pain control.

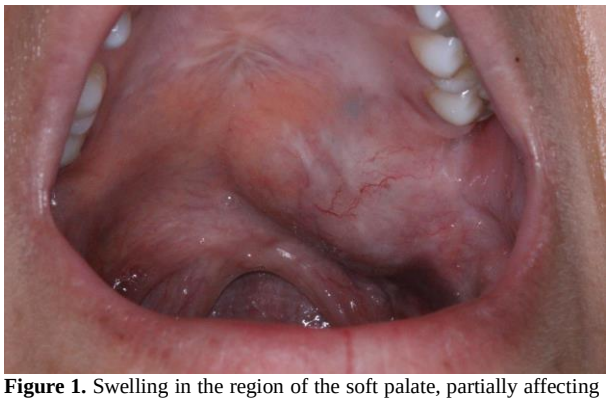

Figure 1. Swelling in the region of the soft palate, partially affecting the nasopharynx.

The area of Oral and Maxillofacial Traumatology Surgery worked in conjunction with Head and Neck Surgery. The lesion was resected with margins of 1 cm on the soft palate and the posterior portion of the hard palate on the left, with the left tonsillary pillar as its limits to the midline (Figure 2), with partial resection of the maxilla and the greater palatine nerve due to tumor involvement, which explained the patient's symptoms (Figure 3). We also performed resection of the posterior nasal spine and left cervicotomy for selective lymphadenectomy at the same surgical time. The resected materials were sent for anatomopathological examination that found the presence of unifocal adenoid cystic carcinoma with compromised margin and perineural invasion, without lymph node metástases (Figure 4).

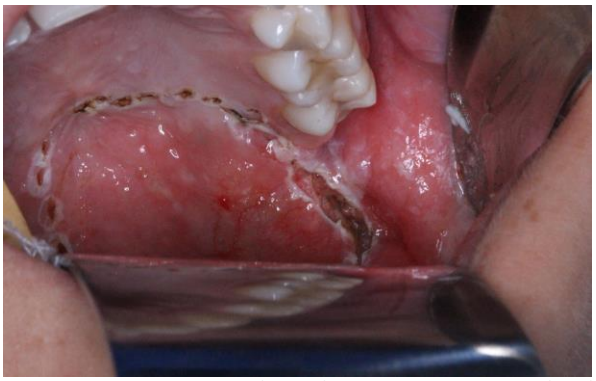

Figure 2. Surgical margin of 1cm from the lesion on the left soft palate, crossing the midline.

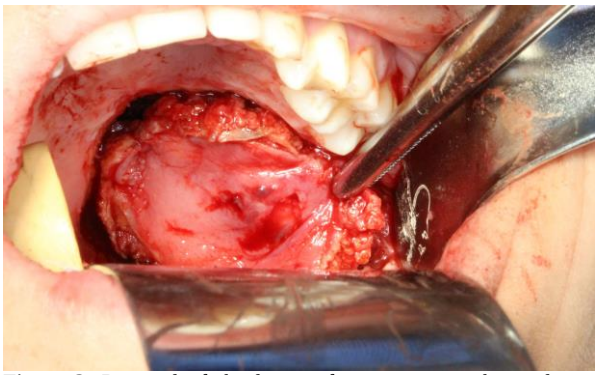

Figure 3. Removal of the lesion after resection with an electric scalpel.

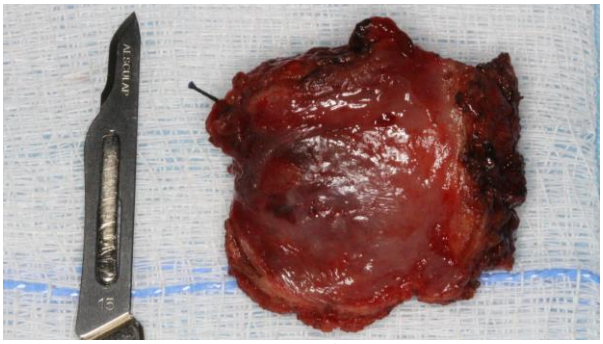

Figure 4. Complete lesion removed after tying and bleeding control, sent for analysis.

The patient was then referred for adjuvant radiotherapy and maintained outpatient follow-up with the Head and Neck Surgery and Oral and Maxillofacial Surgery teams.

Postoperative radiotherapy constitutes the main

negative factor for the quality of life in patients rehabilitated by obturator prosthesis after a maxillectomy, where patients with malignant tumors and who underwent adjuvant radiotherapy can evolve with significant trismus, difficulty in inserting the obturator, dryness oral and mucosal wounds[9]. The obturator prosthesis, in turn, was made from a preoperative plaster study model for the occlusion of the surgical site (Figure 5). It is the main method for the rehabilitation of large maxillary defects. Its main objective is the occlusion of the defect, separating the oral cavity from the nasal cavity, thus preventing nasal speech, nasal regurgitation of food and liquids, in addition to supporting the facial profile[10]. Due to several prosthetic options found on the market, the choice, on the part of the surgeon in conjunction with the prosthodontists, should be based on the location and dimensions of the defect, health conditions, and remaining teeth and bones, available soft tissue reserve and muscle control[11].

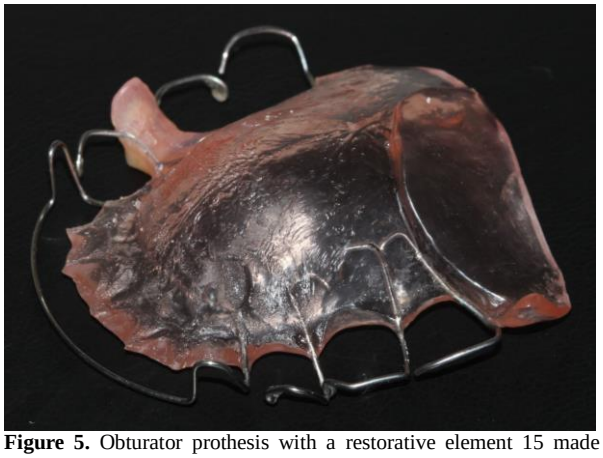

Figure 5. Obturator prothesis with a restorative element 15 made from a preoperative plaster

The patient remained in outpatient follow-up in the postoperative period, weekly, until the second month, after which she was followed up monthly for eight months, presenting a good aspect of tissue repair, but complaining of pain in the surgical site and dysphonia. For this, an analgesic prescription was made and sent for follow-up with the Speech Therapy team.

The treatment of the clinical case proved to be effective, including the esthetic-functional restoration of the patient.

3. DISCUSSION

Adenoid cystic carcinoma is one of the most common salivary neoplasms and occurs most commonly in minor salivary glands, causing swelling of the affected region, pain and a slight predilection for the female sex[12]. These tumors are usually present in young and middle-aged adult patients, with a rare appearance in patients under 20 years of age. They are malignant tumors, which can generate recurrences in a multifocal pattern and with a propensity to generate distant metastases in 40% of the cases, focusing on the lungs and bones, however, this percentage can reach 70% in cases of terminal stages of the disease[13]. Also, tumors on the palate or in the maxillary sinus can eventually invade the base of the skull and spread to the brain[14].

Berg *et al.* (1968), drew attention in his article where he suggested the association of ACC with metastasis in ipsilateral breast, where 7 of his 396 patients had cancer in that region. This association was reaffirmed by Prior and Waterhouse in 1972, who reported that 7 of their patients had an association of breast cancer with neoplasms in the major salivary gland. Years earlier, Shafer & Muhler (1956), had already raised this theory, based on the evidence that hormonal changes from estrogen cause changes in the breast as well as in the salivary glands. His research in rats indicated an etiological role for this hormone at various stages of reproductive life. However, Heap & Broad (1974), stated that the hormonal role of estrogen is not decisive in altering the metabolism of cells and does not represent a significant factor in the development of salivary neoplasms, indicating that further studies in this area would be necessary for such a statement, but reinforce that patients females with both breast cancer and tumors in the salivary glands should be considered malignant until proven otherwise[15].

In 1979, Spiro in his research with other collaborators noticed that there is better control of adenoid cystic carcinoma originating from the tongue and palate compared to those originating from the nasal cavity and the maxillary sinus[16].

Although its cause is still uncertain, it is believed that ACC originates from the ducts of the mucus-secreting glands[17], more specifically from cells that can differentiate into epithelial and myoepithelial cells[18]. Adenoid cystic carcinoma represents about 60% of cases of minor salivary glands, with prevalence in the palatal region. They are known to appear in both minor and major salivary glands (25% in the parotid, 15% in the submandibular and 1% in the sublingual), maxillary sinus, palate, adipose tissue, skin and nerves, which can cause facial paralysis[19,20]. Patients notice a submucosal growth of the lesion, which may be ulcerated or not, with pain as the most common and important symptom in the early stage of the lesion. Upon palpation, these nodules range from hard to soft with possible fluctuation points. Microscopy shows the tumor with cells organized in a swirl around the nerve bundle, which may correspond to patients' pain complaints[21]. Sometimes, associated with adenoid cystic carcinoma, other malignant neoplasms of salivary glands can also be seen, especially low-grade polymorphic adenocarcinoma[22].

William Kuhel (1992), from his clinical research, analyzed 41 patients (22 women and 19 men) who obtained definitive treatment for adenoid cystic carcinoma (all coming from the palate region), dividing them into three categories, depending on its local extent and growth. The ages varied between 20 and 82 years, with an average of 54 years. Of the total number

of patients, 38 tumors were found in the region of the hard and mucous palate, 2 in the soft palate and 1 in the union between hard and soft palate. All patients were treated with surgery and 22 with adjuvant radiotherapy. The category 1 group (C1) presented ACC only in the palate region; category 2 (C2) included tumors that had tumor infiltration in regions such as a nasal cavity, maxillary sinus or pterygoid structures. Finally, category 3 (C3) referred to larger tumors, but clinically limited and with microscopic infiltration far beyond their margins. The analysis of the latter category typically presented tumor cells involving turbinates, nasal mucosa, palate, and pterygoid muscles, in addition to the tumor mass. In C3 patients, younger patients who were prone to early lung metastases were identified, when compared to those in C1 and C2. A total of 4 of the 11 patients in C3 developed metastases over 5 years, from the discovery of the tumor, which is in direct agreement when none of the C1 and C2 patients had metastases. Paradoxically, only 1 patient C3 presented the tumor contained only at the primary site. The authors, from the research, concluded that for patients classified as C1, only surgery with local excision is necessary for treatment, where adjuvant radiotherapy would be necessary only in cases of perineural invasion, with an excellent prognosis. For C2 patients, the authors recommend surgery in addition to radiotherapy on the affected sites. Finally, for patients in group C3, surgery with adjuvant radiotherapy (50Gy to 60Gy) would be necessary in all cases, aiming to decrease relapse in the affected primary site. This was confirmed by the authors themselves, where none of the 9 patients treated with the suggested therapy had local recurrences, as well as a lower risk of lung metastases. However, they claim that these metastases are compatible with prolonged tumor survival[23].

Due to a poor prognosis, it is recommended to avoid aggressive and mutilating treatment of large tumors and in cases where metastases have already been detected. In these cases, the survival rate is limited. Although there is a survival rate of around 70%, this percentage declines over the years, reaching a 25% survival rate after 20 years[24].

4. CONCLUSION

Salivary glands can show a diverse range of neoplasms and cellular changes, being a diagnostic challenge for even the most experienced clinician and pathologist. Although the ACC of the minor salivary gland is a very common tumor, its diagnosis should be made with caution. Complete surgical excision with free surgical margins is the key to the successful treatment of these tumors. Besides, clinical and imaging follow-up will determine the tendency for cystic expansion, identifying and categorizing clinicopathologically in the three categories mentioned, ultimately guiding the most appropriate treatment.

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