

Carcinoma Ex-Pleomorphic Adenoma - A Giant Tumor

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Rezumat

Carcinom ex-adenom pleomorf – o tumoră gigant

Adenomul pleomorf este una dintre cele mai frecvente tumori care afectează glanda parotidă. În absența tratamentului tumora crește constant în dimensiuni și se poate maligniza. Este prezentat cazul unei paciente, suferindă de un carcinom ex-adenom pleomorf, cu o evoluție de peste 20 de ani și care a atins dimensiuni impresionante. Această tumoră, după câte știm, se situează pe locul al doilea la nivel mondial ca dimensiune tumorală a carcinoamelor ex-adenom pleomorf și pe locul nouă între tumorile de parotidă îndepărtate chirurgical. Cu toate acestea, caracterul invaziv regional al tumorii, în acest caz particular, a fost foarte limitat, fără a genera diseminare ganglionară sau metastaze.

Cuvinte cheie: carcinom ex adenom pleomorf, tumori de glande salivare, tumori de parotidă

Abstract

Pleomorphic adenoma is one of the most frequent tumors that involve the parotid gland. The tumor constantly increases in dimension if not cured in due time and may become malignant. A case of a patient suffering from a

carcinoma ex-pleomorphic adenoma that had a 20-year evolution and reached impressive dimensions is presented. The tumor holds the second place worldwide among the largest carcinoma ex-pleomorphic adenoma in terms of size and the ninth place worldwide among the most voluminous parotid tumors ever surgically excised, as far as we know. Nevertheless, the regional invading character of the tumor in this particular case has been limited, without generating local lymph node invasion or metastases.

Key words: carcinoma ex-pleomorphic adenoma, salivary gland tumors, parotid tumors

Introduction

The parotid gland region represents a complex anatomic space that can host various tumors with diverse histological characteristics and clinically diverse symptoms, raising problems of therapeutic management.

Salivary gland neoplasms represent 3-4% of the head and neck tumors. Their incidence reaches approximately 2.5 - 3 cases for 100000 persons each year (1). The pleomorphic adenoma is a benign tumor that may become malignant if untreated for a long period of time causing the emergence of carcinoma ex-pleomorphic adenoma (2). This tumor represents less than 4% of malignant salivary gland tumors and 11.6% of all carcinomas (3).

This paper aims to present a clinical case of a patient with carcinoma ex-pleomorphic adenoma with long term evolution, which reached remarkable size.

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Case report

An 81-year-old female patient with a giant pediculed tumor in the right parotid and masseter region, pendulating to humeral level, addressed our department. The posture of the patient, due to the weight of the tumor mass was in anterior flexion of the thorax on the abdominal region; the patient was not able to hold her head up high unless helped.

Spontaneous self-limited bleedings from the surface of the tumor and the exulceration with the elimination of fetid blood-purulent residues were the reasons why the patient addressed our department. The tumor developed about 20 years ago and had a slow growth in volume ever since. The fear of surgical treatment was the reason for not visiting any physician sooner.

In the past year or so the general condition had gradually started to alter, the daily activities being reduced to resting in bed and finally determined the appearance of decubitus lesions and of necrosis areas on the posterior side of the tumor. The patient presented herself for treatment because of the fetidity of the secretions of the ulcerous area.

Clinical and imagistic examinations

The physical examination revealed the presence of a tumor with the circumference on the implantation basis of 36 cm and the length of 26 cm with the implanting basis at the level of pre-auricular, masseter, parotid, cheek, submandibular and right superior cervical region. In anterior-posterior direction, the tumor spread from the anterior margin of the sternocleidomastoid muscle up to the level of the right angle of the mouth. The tumor presented multiple dilatations with deep epitheliant grooves, large dilated veins, at the posterior pole having two ulcerous pits of 8 to 10 cm in depth (Fig.1), wherefrom a fetid bloody biologic product was expressed.

The general status of the patient was visibly altered as she was presenting real difficulties in performing daily chores. In addition, she was suffering from second degree hypertension, chronic ischemic cardiopathy, chronic cholecystitis, diseases under treatment prescribed by her general practitioner, but the patient followed the medical prescriptions according to recommendations only when clinical evolution got worse.

CT examination with contrast substance was performed in order to evaluate the extension and depth of the tumor, as well as its relations with proximal anatomical formations, especially with the latero-cervical vascular bundle. Thus, a multilobulated structure with areas of necrosis and intra-tumoral calcifications, having a deep polycyclic contour with the cleavage plane against the jugular-carotid bundle, which was embedded into tumoral mass (Fig. 2), was revealed. The peri-tumoral bony structures were intact and of normal aspect. There were homo-lateral latero-cervical lymph nodes with dimensions of 1 - 2 cm in diameter.

Intra-surgical aspects

Ablative surgery was performed under general anesthesia, using



Figure 1. Preoperative clinical aspect

a combined approach, at the level of the anterior margin of the sternocleidomastoid muscle, with the opening of the latero-cervical area and the incision of the tumor circumference. Circumferential dissection of the tumor was undergone, thus observing the main vascular pedicle of the tumor located at the posterior margin of the ascending mandibular ramus. We removed the tumor from the large latero-cervical blood vessels and lifted it as a whole including the parotid gland and cervico-facial ramus of the facial nerve as it was incorporated into and

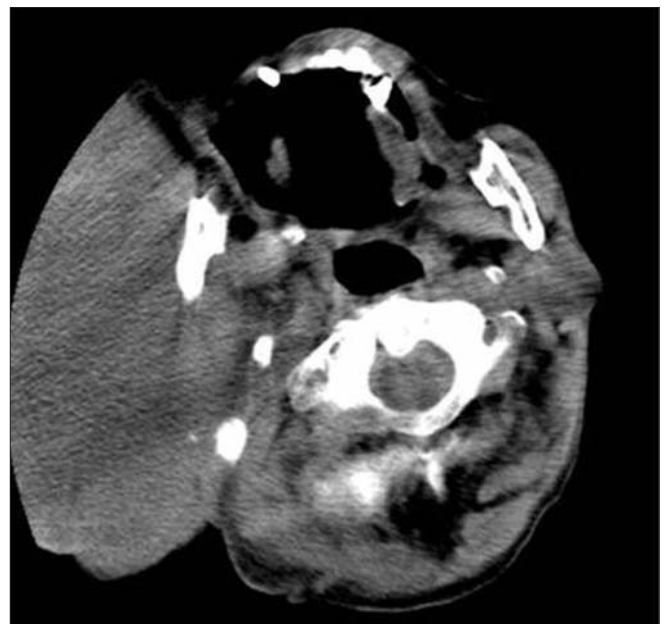


Figure 2. Aspect on CT scan

adherent to the tumor mass. We associated type III radical modified neck dissection, on the right latero-cervical side. Subsequently, the closure of the postoperative defect was performed using nearby random vascularized skin flaps (Fig. 3).

Histopathological examination

Macroscopic post-surgery examination showed a whitish aspect on the section with cysts, necrosis areas, calcifications and focal bleedings. The tumor mass weighted a little over 4300 g and the dimensions were of 26/22/17cm in diameters (Fig. 4).

The microscopic aspect revealed a mixed tumor, carcinoma ex-pleomorphic adenoma. Thus, vast cellular areas specific to pleomorphic adenoma were detected, and centrally, areas of smaller sizes in which cells strongly eosinophilic presenting pleomorphic nuclei and prominent nucleoli were identified, the latter suggesting malign degeneration of certain areas of the tumor of the carcinoma type. Microscopic examination of the lateral-cervical lymph nodes in the neck dissection specimen did not confirm characteristic malignant modifications but a lymphatic stasis with follicular reaction was detected.

Postsurgical evolution

Evolution of the patient after surgery was uneventful, the patient being discharged 7 days after the surgical intervention. The patient returned for examination every two months during the first six months after surgery and then every six months after the first half year. There were no signs of tumor recurrence, adenopathies or metastases. Three years after the surgical ablation of the tumor, the patient ceased to undergo periodical examinations and all contact with our clinic.

Discussion

Parotid tumors may reach, after a long evolution, significant dimensions. According to an international classification of the most voluminous parotid tumors, the current case holds the ninth place, the largest tumor being that case of a 63-year-old patient who had a tumor weighting 26.5 kg, operated on by Frylinck in 1956 (4). As far as we know, if we consider the morpho-pathological features of the tumor, our case holds the second place worldwide, as regards to its dimensions, after the case presented by Honda-Yamamoto in 2005 (5), among malignantly degenerated parotid tumors. The authors are aware of the limits specific to these classifications and of the fact that not all the cases presenting surgically ablated gigantic parotid tumors are published. Nevertheless, there is a moral duty which each member of the medical community ought to have, that is to present the cases which can contribute to this worldwide classification aging 140 years, made in 1989 by Schultz-Coulon (6).

The pleomorphic ex-adenoma carcinoma is preponderantly specific to women in their sixth and seventh decade of age, most frequently at the level of the parotid gland (7). In order to classify a tumor accordingly, areas of pleomorphic adenoma and



Figure 3. Postoperative clinical aspect

of malignant degeneration must be present, as well as the past existence of an excised pleomorphic adenoma that recurred under malignant form. From this point of view, the case presented in this paper can be included in the first category. The malignant degeneration of the tumor occurred at a variable period of time since its onset.

If during its evolution, the malignant component of the pleomorphic ex-adenoma carcinoma exceeds the capsule represented by the benign component of the tumor, this is considered as being an invasive, malignant tumor and thus, there is a risk of metastases in the regional lymph nodes. Brandwein et al (8). did not find metastases or recurrences in cases of tumors which invaded an area smaller than 1.5 mm.



Figure 4. Aspect of excised tumoural formation

However, Felix A et al (9). presented the case of a pleomorphic ex-adenoma carcinoma completely encapsulated, joined by regional metastases within a 10-month-period since the onset of the disease. In the case presented in this paper, the absence of the regional metastases suggests the fact that the evolution of the tumor strictly under the benign form was a long term one, fact that limited the extension of the malignant component, containing the malignancy into the benign component which bordered it. On the other hand, the lymphatic stasis occurring as a consequence of the increase in dimension of the tumor limited the mobilization of the malignant tumoral cells and, implicitly, the occurrence of metastases.

The presented clinical-case confirmed the general opinion of experts according to which benign tumors of the parotid have, sooner or later, the tendency of malignization.

The malignant transformation is independent of the age or the dimensions of the tumor; however, it can be noticed that the appearance of the malignant component after a longer benign evolution, after the tumor had reached considerable size, reduced the invasive nature of the malignant component.

Conflict of interest

There is no conflict of interest.

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