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Malignant Granular Cell Tumor of the Mandible: A Case Report and Review of Literature

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Abstract **Case Report**

The granular cell tumor, also known as Abrikossoff's tumor, is a rare entity. It was first described by Abrikossoff in 1926. It is a ubiquitous site tumor with a predilection for the cervico-facial region. Their pathogenesis has long been debated. After initially proposing a striated muscle origin, recent studies are in favor of a Schwannian neurogenic origin. Malignant forms have been reported in the literature, with mainly lymph node and pulmonary metastases. We report a case of a malignant granular cell tumor of mandibular localization.

Keywords: Granular cell tumor, Malignant, necrosis, mitoses, prognosis.

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INTRODUCTION

The granular cell tumor, also known as Abrikossoff's tumor, is a rare entity. It was first described by the Russian pathologist, Alexeï Ivanovich Abrikossoff in 1926, who named it granulocellular myoblastoma. It is a rare, usually benign tumor, the histogenesis of which has long been controversial. The muscle origin initially suggested was discarded in favor of a nervous origin, or more precisely Schwannian. It is a ubiquitous site tumor with a predilection for the tongue in its intraoral location [1]. Particularly aggressive malignant forms represent less than 2% of cases, with a poor prognosis. The definitive diagnosis is exclusively histological and immunohistochemical.

OBSERVATION

An 82-year-old woman, with no particular pathological history, presented with a budding tumor of the oral cavity which had progressed for one year, initially located at the level of the left mandibular angle and gradually increasing in size. Clinical examination revealed facial asymmetry at the expense of the left hemiface in relation to a hard, painless, cheeky swelling with a smooth surface, fixed in relation to the deep and superficial planes. There was a limitation of the mouth opening to 25mm. The endobucal examination revealed an ulcerative swelling mass, painless, bleeding on contact, infiltrating the labial vestibule beyond the midline, the floor of the mouth in its anterior part without affecting the ventral aspect of the tongue, the internal aspect of the cheek up to 'at the anterior pillar of the palatal tonsil (Figures 1 and 2). Bilateral cervical lymphadenopathy was noted. The general condition was altered with a performance index (OMS) of 3.

The cervico-facial CT scan was in favor of a tumor process encompassing the mandibular ramus with significant bone lysis, extended to the infratemporal fossa, coming into contact with the lateral wall of the pharynx and infiltrating the prestylar region. There was an endocranial extension of the tumor as well as multiple cervical Lymphadenopathy (Figures 3,4 and 5). The thoraco-abdominal scan revealed the presence of multiple secondary pulmonary nodules. A biopsy was taken. Histological examination showed undifferentiated malignant tumor proliferation, made up of large cells with a vesicular nucleus, most often in mitosis. It carries out syncital, isolated clumps and trabeculae. The stroma was fibrous and inflammatory with numerous osteoclastic-like giant cells. Tumor cell immunostaining was positive for anti-S-100 protein, and anti-CD68. Which led to the diagnosis of malignant granular cell tumor. In view of the advanced stage of the tumor, the patient was referred to oncology for palliative chemotherapy.

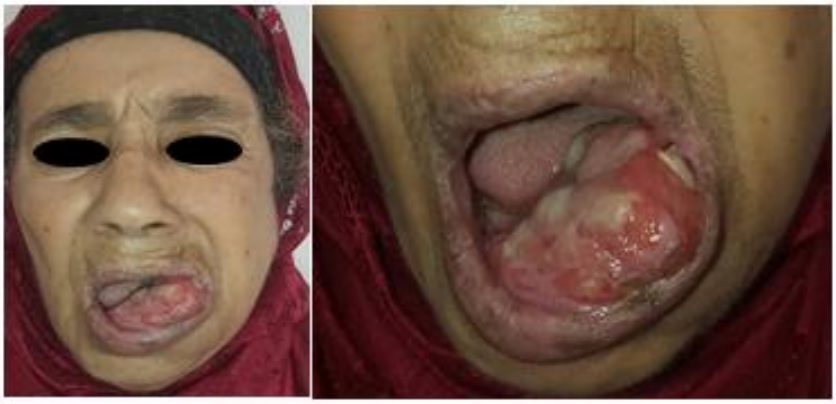

Fig-1, 2: budding tumor of the oral cavity

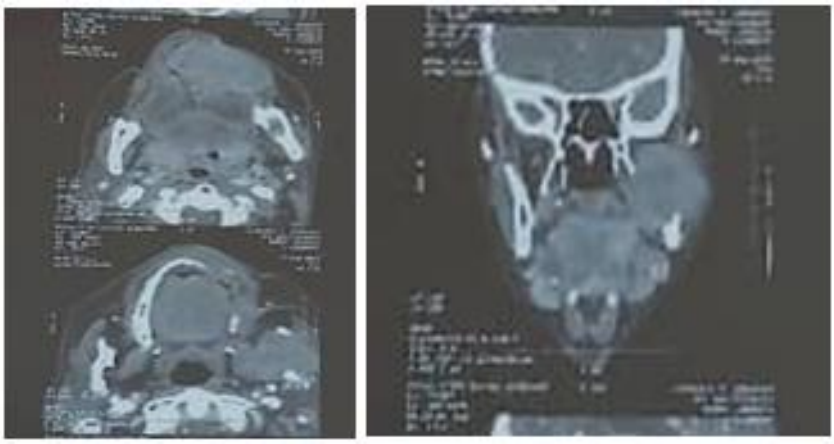

Fig-3, 4: Cervico-facial CT (coronal and axial slices): left mandibular tumor process in the infratemporal fossa

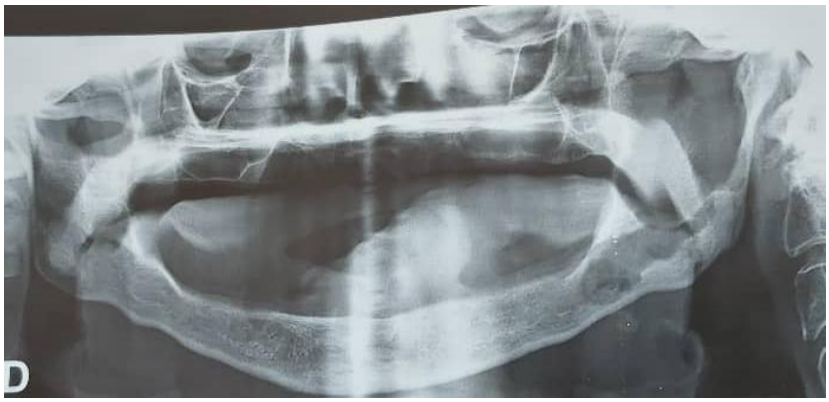

Fig-4: Bone lysis of the ramus and the left mandibular angle

DISCUSSION

The malignant variant of Abrikossof's tumor accounts for less than 2% of all granular cell tumors. It is a rare histological entity [2]. It occurs in adults 20 to 60 years old, with a peak in frequency between the fourth and sixth decades with a clear predominance of women. It is rare in children. It is about twice as common in women as in men and in black people [3]. The preferred location is the cervico-facial sphere [4, 5]. However, a few cases of genital, pulmonary, biliary, laryngeal, and mammary and limb locations have been reported in the literature [6-8]. It most often occurs de novo with, however, the possibility of malignant transformation of a benign form; hence the importance of annual monitoring.

Malignant granular cell tumors mainly described in deep visceral locations or in soft tissues. However, they can have a cutaneous or mucous membrane localization. They are clinically suspected in the presence of a lesion larger than 4cm in size, the existence of necrotic and / or hemorrhagic areas as well as rapid growth. Because of its clinical polymorphism, only a histological examination coupled with an immunohiscochemical study can confirm the diagnosis. Epidermal hyperplasia, PAS cytoplasm positivity, S100 protein positivity are common to both benign and

malignant forms. However, given the often atypical nature of the lesion, several diagnostic classifications have been issued. Fanburg-Smith et al. developed a more precise classification with six criteria (tumor necrosis, spindle cells, vesicular nucleus with large nucleoli, mitotic index greater than 2 for ten fields, high nucleocytoplasmic ratio and pleomorphism) The presence of at least three of these criteria makes it possible to consider the tumor to be malignant [6, 9, 10]. Three of the criteria were found in our case, in addition to the extensive and progressive nature of the tumor process as well as the presence of metastatic pulmonary lesions. Proliferative activity is assessed by the expression of Ki 67 and P 53. A Ki 67 level greater than 10% is considered by some authors to be an unfavorable prognostic factor.

The differential diagnosis is mainly made with melanoma, malignant tumor of the peripheral sheath, dermatofibrosarcoma, sarcomatoid carcinoma, leiomyosarcoma and angiosarcoma. Hence the interest of performing immunostains, such as Melan-A, Smooth Muscle Actin, CD34, CD31, HMB-45, cytokeratin, CD68 and S100 is highly recommended.

Due to the unfavorable prognosis of the metastatic forms, the malignant forms require an extension assessment to look for secondary lymphatic or systemic locations (lung, liver, bone). Indeed, secondary pulmonary locations are frequently described, and locoregional lymph node invasion accompanies most malignant tissue lesions. This was the case with our patient.

The treatment of choice for malignant granular cell tumors remains surgical excision. Due to its non-encapsulated and infiltrating character, good margins are recommended. Many authors report incomplete resections; in the reported case, the excision brushed the deep edge of the tumor and the lesion, not encapsulated, penetrated deep into the muscles of the tongue [1,8]. Thus some authors recommend excision margins of 2 to 3 cm. However, in the oral cavity, tissue preservation is essential; Moh's micrographic surgery may then be indicated. Both adjuvant radiotherapy and chemotherapy have not been shown to be effective but should be considered in inoperable patients.

The prognosis is relatively poor, with a high local recurrence rate (70% and an average survival of 45.1 months).

CONCLUSION

Although rare, malignant granular cell tumor is an aggressive tumor with a poor prognosis, hence the benefit of early diagnosis and management based on surgical excision with safety margins varying from 2 to 3 cm. Due to the recurrent and metastatic potential, rigorous annual monitoring is required.

Contributions of the authors

All the authors contributed to the medical care of the patient, as well as the writing this article they approved.

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