

Abrikossoff's tumor of the tongue: Report of three cases and review of the literature

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Abstract

Abrikossoff's tumor, also called granular cell tumor (GCT), is a rare benign neoplasm of the soft tissues. In almost 70% of the cases, it occurs in the head-and-neck region (especially in the tongue), even though it may present in every other part of the body. This neoplasm has a benign behavior usually, but there have been described a malignant transformation in 2%–3% of the cases. The characterization of this tumor depends on its clinical and histopathological findings. The purpose of our work is to report three uncommon cases of oral Abrikossoff's tumor located on the tongue in young patients referred to our department of maxillofacial surgery of "University Magna Graecia" in Catanzaro, also reviewing of the literature.

Keywords: Abrikossoff's tumor, granular cell tumor, nodular lesion, tongue lesion

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INTRODUCTION

First, described in 1926 by Abrikossoff and initially called myoblastoma due to its suspected muscle fibers origins, this tumor appears to be a benign soft-tissue neoplasm of unknown etiology, but multifocal lesions have also been described.^[1] It is usually an asymptomatic solitary sessile nodule, well-circumscribed with typical pink overlying mucosa and slowly growing. It can involve any part of the body, but in most cases, it localizes in the head-and-neck region, especially the tongue. It is more common between the fourth and the sixth decade, very few cases of earlier presentation (younger than 20 years old) have been described.^[2] Women are affected

twice more than men, and the black ethnic groups are more interested than the Caucasian population. The differential diagnosis of granular cell tumor (GCT) of the tongue includes numerous benign mesenchymal tumors such as neurofibroma, vascular lesions of the tongue, pleomorphic adenoma of the minor salivary glands, dermoid cyst, lipoma, traumatic fibroma, neuroma and schwannoma.^[3] The classical treatment of Abrikossoff's tumor is surgical excision with safety margins although this is not always possible because tumor can lack the capsule. Here, we report three cases firstly diagnosed through biopsy as GCT in a very young population.

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CASE REPORTS

Case 1

A 25-year-old male presented at our department with a painless nodular lesion incidentally discovered 5 months earlier. Clinical examination showed a right-seated mass, well-defined, non-tender and non-fluctuant, involving the body of the tongue on the right side and measuring 1.5 cm × 1 cm [Figure 1]. Ultrasound examination of the tongue and neck has been performed, and they revealed reactive inflammatory lymph nodes excluding a malignant disease. The patient appeared to be in the overall good health. He underwent incisional biopsy of the mass reporting diagnosis of “granular cell tumor.” Subsequently, a magnetic resonance imaging examination (MRI) was demanded, which reported “a soft-tissue mass in the right side of the tongue showing contrast enhancement in T1 sequences” [Figure 2]. The tumor was totally excised under general anesthesia, and histopathological examination confirmed the biopsy diagnosis. In particular, hematoxylin and eosin-stained sections revealed pseudoepitheliomatous hyperplasia of the overlying stratified squamous epithelium, circumscribing proliferation of monomorphic elements with abundant granular cytoplasm [Figure 3]. No mitotic figures were observed. At immunohistochemistry, all neoplastic cells were S-100 and C68 positive, Ki67 <10% [Figure 4]. Surgical margins were free from disease. All these findings confirmed the diagnosis of Abrikossoff's tumor of the tongue. The patient was first examined 1 week later and again with a regular follow-up after 1, 3 and 6 months [Figure 5]. So far, no sign of recurrence has been noted, as highlighted by postoperative ultrasound and magnetic resonance examination [Figure 6].

Case 2

A 19-year-old Caucasian female I.C. presented at our department with painless lingual swelling, arose approximately in the past year. An intra-oral examination revealed a nodular, sessile lesion, well limited, of hard consistency, whose texture was similar to that of adjacent mucosa. The nodule of roundish morphology was located on the right side of the dorsum of the tongue near the apex, 2 cm in diameter [Figure 7]. Initial imaging study with MRI was prescribed by general practitioner, reported a “pseudonodular area of 1.4 cm × 0.7 cm × 1 cm, hypointense on T1 sequences and contrast uptaking on lesion.” An incisional biopsy was performed, and the histopathological diagnosis was Abrikossoff's tumor; so, the patient was treated under general anesthesia for a complete lesion excision. The histopathological examination confirmed the biopsy diagnosis; in particular,



Figure 1: Case 1. Abrikossoff's tumor on the dorsum of the tongue



Figure 2: Preoperative magnetic resonance imaging showing lesion with contrast enhancement in T1 sequences

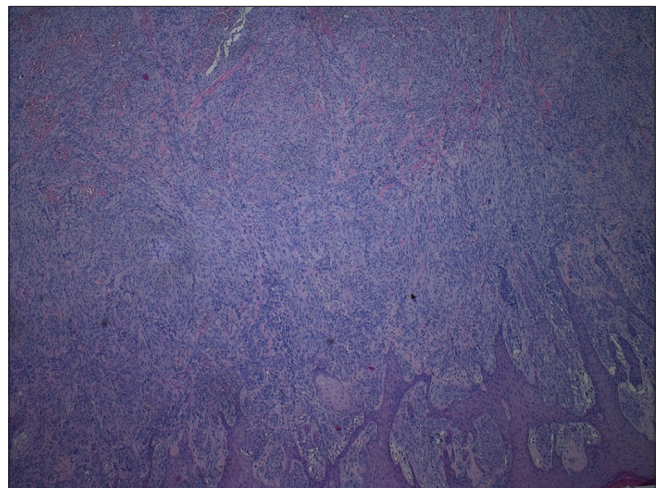


Figure 3: Hematoxylin and eosin section showed pseudoepitheliomatous hyperplasia (magnification, ×5)

the microscopical analysis showed pseudoepitheliomatous hyperplasia. At immunohistochemistry, the specimen resulted in S-100 positive and CD68 positive. The

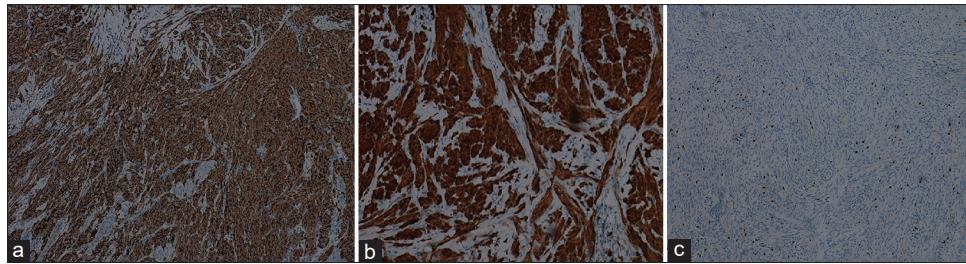


Figure 4: Histopathological examination reveals that all neoplastic cells were C68 (a) and S-100 (b) and positive, Ki67 <10% (c) (magnification, ×10)



Figure 5: Postoperative clinical evaluation after 6 months

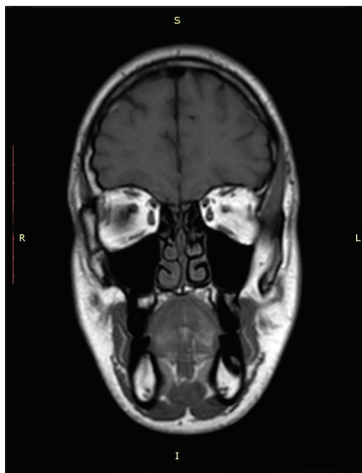


Figure 6: Postoperative magnetic resonance imaging scan showing complete excision of tumor

patient was first examined 1 week later and with a regular follow-up after 1, 3 and 6 months after the surgical excision with no clinical and radiological signs of local recurrence.

Case 3

A 32-year-old male came to our department for an early arose tongue mass. The tumor appeared as well-circumscribed mass, covered by the healthy mucosa,

<2 cm of major diameter, unpainful, located at the left margin of the tongue [Figure 8]. An incisional biopsy was performed, which diagnosed “Abrikossoff’s tumor of the tongue.” Unfortunately, the patient refused more than once, the surgical excision due to personal reasons, and he is already subjected to regular follow-up visits to check the behavior of the mass.

DISCUSSION AND LITERATURE REVIEW

Abrikossoff’s tumor or GCT is a rare benign neoplasm commonly observed in the head-and-neck region, especially the oral cavity.^[4] Its origins are controversial;^[5] even though there may be congenital cases. First, described by the Russian pathologist Alexei Ivanovich Abrikossoff in 1926 in a patient with tongue tumor. At first, he proposed a myogenic origin classifying it as a myoblastoma. In 1931, it was described as granular cell myoblastoma due to the histological similarity with skeletal muscle fibers.

Many theories on the origin of GCT have subsequently been proposed, including its origin from striated muscle, histiocytes, fibroblast, myoepithelium and a neural origin.^[6-8] Nowadays, neural origin or differentiation, in particular from Schwann cell type, is the most supported. This is probably based on the close anatomical relationship of GCTs to peripheral nerve fibers, on the ultrastructural demonstration of myelin figures and axon-like structures, and on immunohistochemical reactivity with S-100 protein, neuron-specific enolase and myelin proteins.^[9] The benign GCTs are, in most reports, specifically S-100 and/or neuron enolase positive. Granular cells are strongly positive for the S-100 protein, but there are studies reporting GCTs located in lips and neck, whose cells are negative for that protein.^[10] Neurogenic origin is confirmed in a series of GCT cases with positivity of Vimentin, CD-68, S-100, PGP9.5 and NSE. An unusual and significant microscopic finding is the presence of acanthosis or pseudocarcinomatous hyperplasia of the overlying epithelium, reported in up to 50% of all cases. This hyperplasia in some cases, it may be so pronounced that leads to a mistaken diagnosis of



Figure 7: Case 2. Preoperative imaging of Abrikossoff's tumor on the dorsum of the tongue



Figure 8: Case 3. Preoperative imaging of Abrikossoff's tumor on the left margin of the tongue

squamous cell carcinoma and subsequent unnecessary cancer surgery.

A study of Regezi *et al.* indicates great antigen heterogeneity in this tumor.^[11] In some cases, Fine and Li demonstrated the positivity of inhibin alpha-subunit and calretinin.^[12]

GCT occurs generally in the fourth to sixth decades of life, while in the pediatric population, it is almost rare. Epidemiology studies demonstrate that women are more affected than men, statistically twice as much. Abrikossoff's tumor can affect any part of the body; the most common site in 70% cases is the oral cavity, especially the tongue, where it usually rises in the dorsal surface, but cases involving cheek mucosa, hard palate, lip, gingiva, uvula and parotid gland have also been described even though with less frequency. The tumor is also seen in the skin, gastrointestinal tract, respiratory tract, nervous system and male and female reproductive system. Clinically, GCT of the oral cavity appears to be as a single, smooth nodular mass of pink color with <2 cm in diameter. It is asymptomatic, hard in consistency, covered by intact overlying mucosa. On the tongue, 48% of lesion occur in the dorsum, 15% on the lateral border and the rest on the ventral surface.^[13-15] Diagnosis may be difficult because it includes a wide range of benign connective and neural tumors such as fibromas, lipomas, neuromas, neurofibromas and pleomorphic adenoma of the minor salivary glands, which must be considered in the differential diagnosis.^[7]

Immunohistochemical analysis reveals positivity for S-100 protein within the cells – a finding that is supportive, but not diagnostic, of neural origin. The lesional cells also

are positive for CD-68, calretinin and neuron-specific enolase. An unusual and significant microscopic finding is the presence of acanthosis or pseudocarcinomatous hyperplasia of the overlying epithelium, reported in up to 50% of all cases. This hyperplasia in some cases, it may be so pronounced that leads to a mistaken diagnosis of squamous cell carcinoma and subsequent unnecessary cancer surgery. This tumor, in most cases, is a benign lesion, but only a few cases, in which it turned to be malignant, have been found in literature. The malignant Abrikossoff's tumors have two distinct variations: the first one has benign histopathology, even if characterized by increased mitotic activity and mild nuclear pleomorphism. The clinical features of large size, rapid growth and surface ulceration must, therefore, be used to arrive at a malignant diagnosis, and the pathologist should carefully evaluate the lesional periphery for signs of a true invasion. The second variant shows the transition from typical benign granular cells to pleomorphic granular cells to pleomorphic nongranular spindle cells and giant cells with numerous mitotic figures. Many histologic features, such as necrosis, spindling, vesicular nuclei with prominent nucleoli, more than 2 mitoses/10 HPF, high nucleo-cytoplasmic ratio and pleomorphism, may aware us for an increased risk for metastasis. Tumors with three or more of these features are considered malignant and have approximately 40% risk of causing death.

The diagnostic confirmation is histopathological, but magnetic resonance or computed tomography with contrast medium is fundamental to the correct localization of the mass.

Conservative excision is the treatment of choice for GCT.

CONCLUSIONS

The number of reported cases of Abrikossoff's tumor in literature is low, due to the rarity of the tumor (reported prevalence between 0.019% and 0.03% of all human neoplasms). Despite its low prevalence, GCT should be considered in the differential diagnosis of oral lesions, particularly when located in the tongue. Even though this tumor is an easily recognizable entity, its biological nature and histogenesis remain controversial. Although the cases of malignant lesion are few, to avoid local recurrence of the tumor, is recommended both a complete surgical excision and an accurate histological examination with immunohistochemical panel; is also needed a close follow up. In our cases, positivity of markers such as CD-68 and S-100 and low Ki-67 proliferation confirmed once again the mentioned theories about neuronal origin and benignity of this tumor, furthermore, the immunohistochemical profile was helpful in determining the clinical behavior of the tumor and establishing the final diagnosis with appropriate treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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