

# Abrikossoff Tumor Of Tongue – A Rare Presentation

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## **Abstract:**

*Granular cell tumor is a rare benign soft tissue tumor that most likely arises from Schwann cells. A 19 year old female reported with nodular swelling of size 1 x 1 cm on the dorsum of the tongue for a period of 3 months. Excision biopsy showed the presence of connective tissue stroma with sheets of polygonal cell having eosinophilic inclusions and pustulovuloid bodies of milan. IHC marker S-100 positive confirmed neural variant of granular cell tumor.*

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## **I. Introduction:**

Granular cell tumors were described by Russian pathologist Abrikossoff in the year 1926 (1). They were initially known as granular cell myoblastomas and believed to have a muscular origin. With the development of immunohistochemistry and electron microscopy, they are now postulated to have a Schwann cell origin. A subset of these tumors are positive for S-100 IHC marker and have been referred to as 'neural variant of granular cell tumors'.

These tumors present as painless solitary nodules and are in most cases reported as an incidental finding. They are more common among females and site predilection vary from skin, oral cavity, GI tract , breast , respiratory , neural and genitourinary tract (2)(3)(4)(5).

They are also found to be associated with syndromes such as Noonan syndrome , neurofibromatosis and LEOPARD syndrome. Although vast majority of these tumors behave benign, 1-2% of cases have shown malignant nature with recurrences and fewer curative options beyond surgical excision(6).

## **II. Case Report**

A 19 year old female reported to OMFS department in Government Dental College Kottayam with complaint of a nodular growth on the dorsum of tongue for a period of 3 months. The growth was solitary, nodular, firm ,non exophytic, non tender, non pruritic lesion of size 1 x 1 cm with no associated bleeding and surface changes. The growth was transillumination negative with dry tap on wide bore aspiration. Patient had complaints of associated halitosis which was correlated to difficulty in maintaining tongue hygiene.

The patient was evaluated for routine blood values and after confirming the systemic wellbeing of the patient , the patient underwent a wide local excision of the lesion under local anaesthesia. The excised specimen was sent for histopathology and IHC evaluation

## **III. Histopathological Evaluation:**

Microscopy of the soft tissue specimen showed parakeratinised stratified squamous epithelium overlying a moderately collagenous connective tissue stroma. Epithelium showed pseudoepitheliomatous hyperplasia. Connective tissue stroma showed polygonal cells with eosinophilic granular cytoplasm and eccentrically placed pale stained vesicular nuclei. Distinct oval or round eosinophilic inclusions surrounded by clear halo that correspond to pustule-ovoid bodies of Milan were noted suggestive of Granular cell tumor IHC marker S-100 was positive suggestive of neural variant of granular cell tumor.

#### IV. Discussion:

Granular cell tumors usually found as a benign solitary slowly growing nodular lesion. They are most commonly seen in skin and soft tissue but also reported to be seen in breast, GI tract and internal organs. The head and neck region account for 40-60% cases with tongue being the most common site of predilection for granular cell tumors of oral cavity(2). They are most commonly seen in dermal, subdermal and submucosal areas and less common in skeletal muscles. There have been reported cases of neural and non-neural variants that are distinguishable based on IHC marker S-100.

These granular cell tumors commonly are painless and benign, however aggressive malignant counterparts have been reported.

Malignant granular cell tumors account for 1-3%cases. These malignant variants present with much larger lesions when compared to benign variant. These malignant lesions are usually associated with metastatic disease more commonly to lung. Fanburg-Smith et al outlined histologic criteria to differentiate between benign, atypical, and malignant granular cell tumors(7).

The histo-morphologic criteria that might help predict malignant behavior included

1. spindling of tumor cells
2. increased nuclear/cytoplasmic ratio
3. vesicular nuclei with large nucleoli
4. pleomorphism
5. necrosis
6. increased mitotic activity (> 2 mitoses/10 high power fields at ×200 magnification)

#### Fanburg Smith criteria:

3 or more criteria	Malignant
1 or 2 criteria	Atypical
None	Benign

In the present case scenario , clinically the growth was well circumscribed and slow growing with no associated tenderness. Histopathological specimen did not show any features suggested by Fanburg – Smith etal of malignancy(7). To check the neural tissue involvement , immunohistochemistry study with S-100 was performed. The result of S-100 marker was positive suggestive of neural variant of granular cell tumor. Other IHC markers such as laminin, neuron specific enolase and myelin proteins have also been reported in other studies(8). Radiological evaluation with MRI images have also been an adjunct is distinguishing malignant granular cell tumors in breast and internal organs.

Wide excision is the preferred treatment for patients with benign and atypical lesions and is generally curative. Recurrence in patients with benign granular cell tumors is rare and usually related to incomplete resection. The role of adjuvant chemotherapy and radiation has limited understanding.

#### V. Conclusion:

Granular cell tumors are rare neoplasms whose clinical and radiologic findings are often indistinguishable from other, more common lesions. These tumors, whether malignant or benign, typically present as solitary masses in a wide range of organ systems without any unique features until examined microscopically. Furthermore, while some of the malignant lesions present as such with nodal or distant metastases, many are not found to be malignant until the histology is worked up. Subsequently, while a single physician may be initiating the care of a patient with a granular cell tumor, it is important to consult with an interdisciplinary team of specialists promptly to expedite workup and treatment.

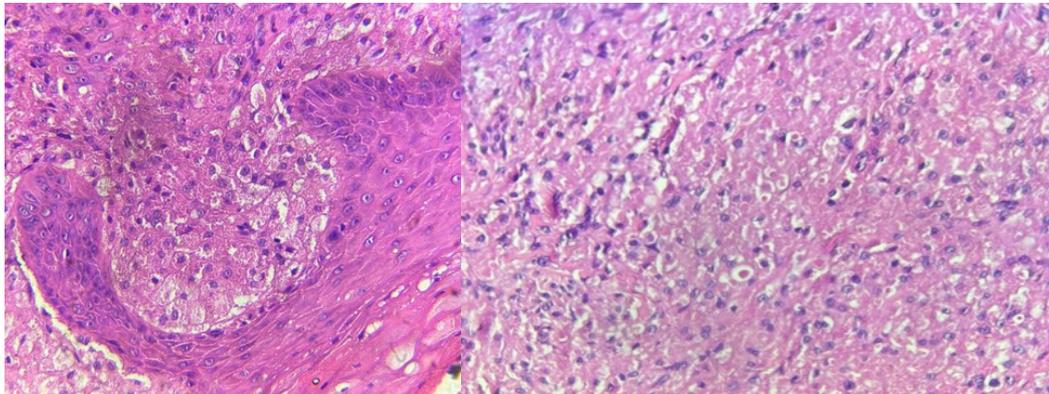
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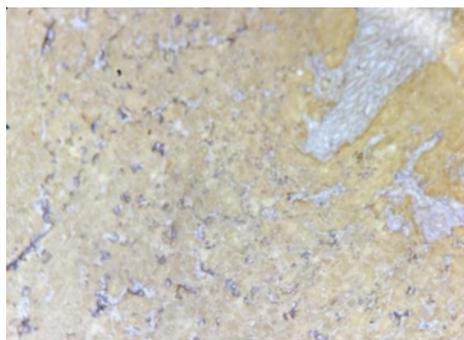
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**Fig 1 Nodular Growth Over Dorsum Of Tongue**  
**Fig 2 Post Excision Healing After A Week**



**Fig 3 Histologic Specimen Showing Parakeratinised Epithelium With Collagenous Stroma With Eosinophilic Inclusions**  
**Fig 4 High Resolotion Specimen Showing Pustule-Ovoid Cells Of Milan With A Clear Halo**



**Fig 5 Immunohistochemistry Showing Positive Stain For S-100 Marker Implying Neural Variant Of Granular Cell Tumor Of Tongue**