# Granular Cell Tumor of the Tongue: A Case Report

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## Abstract:

Granular cell tumor is an uncommon soft tissue tumor; known for its characteristic constituent cells with abundant granular cytoplasm. Recent evidences confirm its neurological origin; however; the etiology remains unknown. The tumor predominantly affects adult population in their $4^{th}$-$6^{th}$ decades; mainly affecting females; and the childhood presentation is unusual. The present case is about a 13 years old Saudi boy with a granular cell tumor in the tongue. The clinical presentation, histological features are described with the review of literature.

**Key Words: C**hildhood presentation, Granular cell tumor, Neurological origin.

## Introduction:

Granular cell tumor (GCT) is an uncommon neoplasm of soft tissues that may occur in any part of the body; however, about 50% of GCTs affect oral cavity and the tongue in particular (Neville *et al.*, 2002; and Billeret, 1999).

GCT is usually presents as a single painless sessile nodule that is either dermal, submucosal or in muscle tissue. Its surface can be smooth or pseudo-ulcerated; and the tumor rarely measures larger than 3 cm in size. Uncommonly in about 10-15% cases, GCT can occurs as multiple nodules appearing synchronously or simultaneously (Billeret, 1999; Argenyi *et al.*, 2006; and Roberto *et al.*, 2011).

Peak incidence of GCT is between $4^{th}$ to $6^{th}$ decades. Females are affected twice more commonly than males while children are rarely affected (Nagaraj *et al.*, 2006).

Initially, the GCT was thought to be of muscle origin and named earlier as the granular cell myoblastoma. However, its common association with the nerves and immunohistochemical properties have firmly established it to be of the neural origin (Billeret, 1999; Nagaraj *et al.*, 2006; and Mazur *et al.*, 1990).

Local excision is the exclusive treatment of choice for GCTs and recurrence is rare (Mazur *et al.*,1990). Malignant GCT was reported in about 2% of all GCT cases (Argenyi *et al.*, 2006).

**Case report:**

A 13 years old, Saudi boy presented to the Oral and Maxillofacial Surgery Department, King Fahad Hospital, Hofuf; complaining of a painless mass in his tongue of one year duration. The mass has been almost stationary in its size without any history of rapid growth or any hindrances in the tongue movements.

There was no family history of any neoplastic conditions. His medical history was unremarkable except for bronchial asthma.

Physical examination was similarly average without any facial deformity or lymphadenopathy.

Intra-oral examination showed a firm non – ulcerated, painless nodule of the dorsum of right side of the tongue measuring 1.5 X 1cm. (Fig. 1). The nodular lesion was well defined with flattened surface and smooth outline. Surgical excision was done under general anesthesia upon the patient's preference (Fig. 2) and the surgical site was closed primarily (Fig.3).

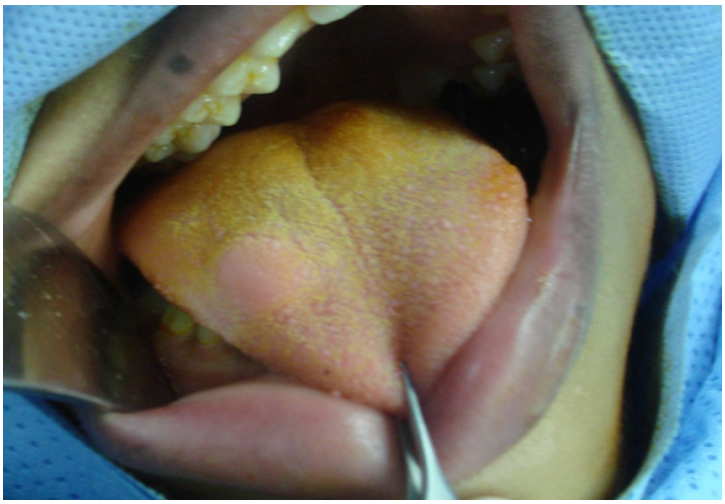

Fig. 1: Intra-oral view shows firm mass of right side of the dorsum of the tongue with loss of papillae of its covering mucosa.

Excised mass was sent for histopathological examination, which revealed the tumor mass to be comprised by diffuse sheets of almost uniform cells with small nuclei and abundant pale pink granular cytoplasm. At many places, the tumor cells contained large coarse particles. The tumor thus conformed to the classical features of granular cell tumor (Fig 4). Any nuclear atypia, nucleoli, necrosis or mitotic

activity was not present.

Patient was followed up for ten months without evidence of recurrence.

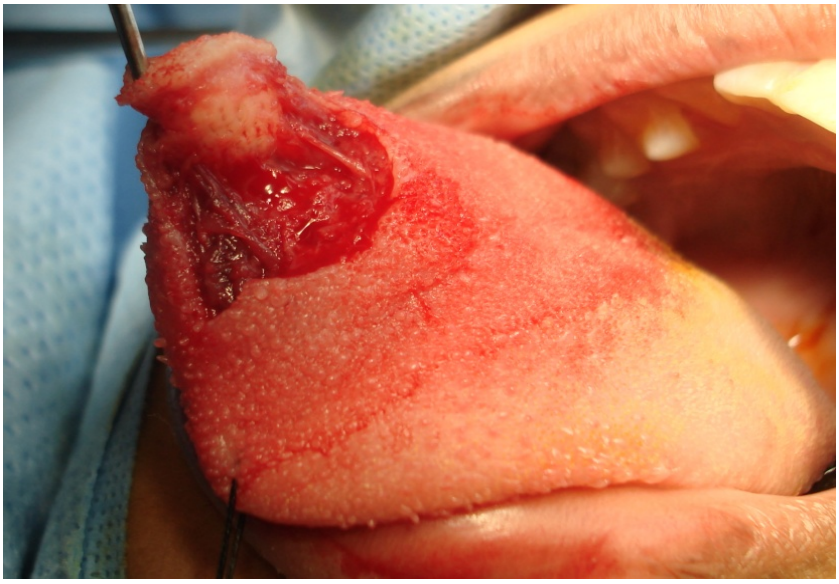

Fig. 2: Intra-operative view showing the excised mass with firmly adherent covering mucosa.

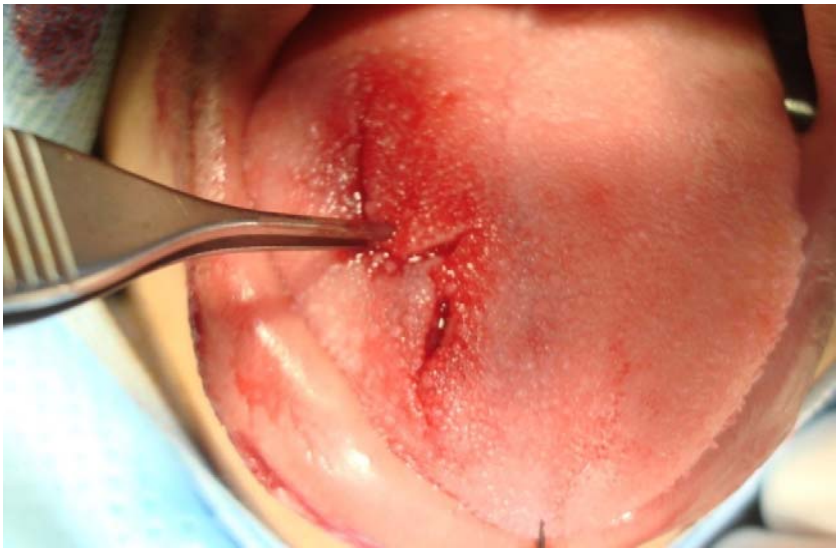

Fig. 3: Intra-operative view shows the approximation of the edges of the excision site which primarily closed.

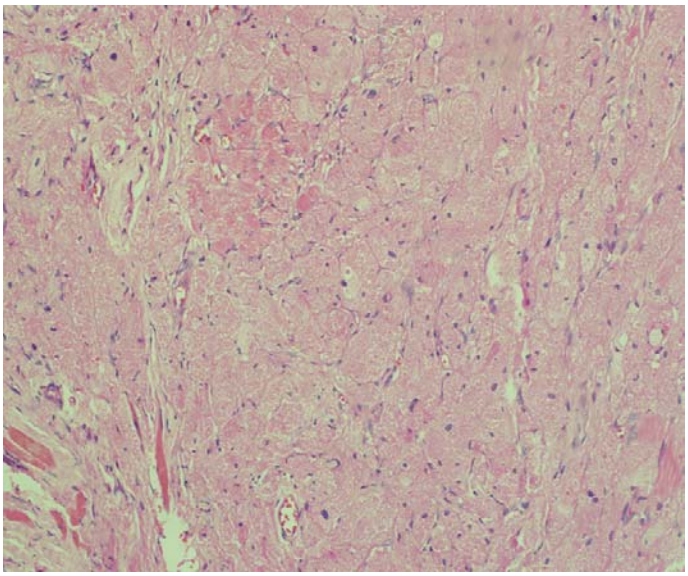

Fig. 4: Microphotograph showing diffusely present large tumor cells with granular cytoplasm (H and E x 40).

**Discussion:**

Granular cell tumor was described first by Abrikossoff as a muscle tumor. Later observations about the frequent granular cell change in the nerves and neural tumors coupled with immunohistochemistry findings for s-100 protein and myelin proteins (PO and P2) in the tumor cells and the ultrastructural evidences strongly support the neural (schwannian) origin of the tumor (Nagaraj *et al.,* 2006; and Sposto *et al.,* 2006).

Clinically, the tumor is presented as an asymptomatic single sessile nodule. GCT occurrence in the tongue represents 23-28% of all head and neck cases (Sposto *et al.,* 2006). It is usually no larger than 3 cm and its surface can be ulcerated. Most of the GCTs behave in benign fashion; rare malignant GCTs are suggested by being painful, larger in size, rapidly growing, recurrent and invading the adjacent structures. Malignant GCTs are more common in extremities than head and neck region including the oral cavity, age and gender predilection is similar to benign ones (Argenyi *et al.,* 2006). Metastatic spread is a common finding in malignant GCTs. (Billeret, 1999; Argenyi *et al.,* 2006; and Apisarnthanarax, 1982). Malignant change in the GCT histologically is indicated by diffuse nuclear atypia,

prominent nucleoli, spindling of tumor cells, mitotic activity (>2 mitoses/10HPF) and necrosis (Neville *et al.,* 2002; Billeret, 1999; Argenyi *et al.,* 2006; and Apisarnthanarax, 1981).

The covering epithelium shows pseudoepitheliomatous hyperplasia of the overlying squamous epithelium in about 50–60% of all cases and up to 87% in a study done by Eguia *et al.* in 2006 (Eguia *et al.,* 2006).

This histological features that mimics infiltrative growth patterns of the islands of squamous epithelium may lead to the lesion being mistaken for a squamous cell carcinoma. However, the coexistence of GCT and true squamous cell laryngeal carcinoma has also been reported (Argenyi *et al.,* 2006; Eguia *et al.,* 2006; Le *et al.,* 2004; and Lassaletta *et al.,* 1998).

The immunohistochemical analysis for oral GCTs shows immunoreactivity for S-100, p75, NSE (neuron specific enolase) and CD-68, and absence of any immunoreactivity for Ki-67, Synaptophysin, SMA (smooth muscle actin), EMA(epithelial membrane antigen), GFAP (Glial fibrillary protein), desmin and Chromogranin. Positivity to s-100 protein is the most used mark for diagnosis of GCTs (Nagaraj *et al.,* 2006; and Vered *et al.,* 2009).

Clinically the differential diagnosis of GCT in the oral cavity may include other connective tissue tumors, such as neuorofibroma, schwannoma, palisaded encapsulated neuroma, lipoma, traumatic fibroma, amyloidosis and other benign mesenchymal neoplasms that may present intraorally as asymptomatic lumps similar to GCTs. The only way to establish a definitive diagnosis is through biopsy (Regezi *et al.,* 2003; and Koloktronis *et al.,* 2003).

Histopathologically, congenital granular cell epulis (CGCE) is very much close look alike of GCT, which occurs in infant exclusively along the labial aspect of dental ridge; here granular cells stain negative for s-100 protein (Basile and Woo, 2003; and Silva *et al.,* 2007).

Surgical excision is the treatment of choice for GCTs; and is considered sufficient, as the recurrence rate is insignificant. However, recurrence and metastasis is quite evident in malignant cases with percentages reach 31-41 % for local recurrence and 50-63% for metastasis (Mazur *et al.,* 1990; Eguia *et al.,* 2006; Giuliani *et al.,* 2004).

This particular case is interesting due to the occurrence of granular cell tumor in a very young male patient; as GCT commonly occurs in the female patients in their 4th-6th decades. We came across a few similar cases, a granular cell tumor in a 14 years Old Italian boy that was also described as

rare (Barbieri *et al.*, 2011). Regionally in Saudi Arabia, a similar case of a 20 years old female in Najran (John and Daniels, 2009). Although rare, such a case suggests that GCT should be included for the possible differential diagnosis even for very young individual.

## References:

Apisarnthanarax, P. 1981. Granular cell tumour: an analysis of 16 cases and review of the literature. J. Am. Acad. Dermatol. 5: 171-82.

Argenyi, Z.B. 2006. Granular cell tumour. In: LeBoit, P.E., Burg G., Weedon D., and Sarasin A. World Health Organization Classification of Tumours. Pathology and Genetics Skin Tumours. IARC Press, Lyon. p. 274-275

Barbieri, M., Musizzano, Y., Boggio, M., and Carcuscia, C. 2011. Granular cell tumour of the tongue in a 14-year-old boy: case report. ACTA Otorhinolaryngologica Italica. 31: 186-189.

Basile, J.R., and Woo, S.B. 2003. Polypoid S-100-negative granular cell tumor of the oral cavity: a case report and review of literature. Oral Surg. Oral Med Oral Pathol Oral Radiol Endod. 96: 70-6.

Billeret, Lebranchu V. 1999. Granular cell tumor. Epidemiology of 263 cases. Arch. Anat. Cytol. Pathol. 47: 26-30

Eguia, A.**,** Uribarri, A., Escoda, C.G., Crovetto, M. A., Martínez-Conde, R., and Aguirre, J.M. 2006. Granular cell tumour: report of 8 intraoral cases. Med. Oral Pathol. Oral Cir. Bucal. 11: E425–E428

Giuliani M., Lajolo C., Pagnoni M., Boari A., and Zannoni G.F. 2004. Granular cell tumor of the tongue (Abrikossoff's tumour). A case report and review of the literature. Minerva Stomatol. 53: 465– 469.

John, S., and Daniels, M. 2009. Granular cell tumour of tongue: A case report. The Saudi Dental Journal. 21: 75–78.

Koloktronis, A., Chatzigiannis, I., and Paloukidou, N. 2003. Oral involvement in a case of AA amyloidosis. Oral Dis: 9: 269-272

Lassaletta, L., Alonso, S., Granell, J., Ballestin, C., Serrano, A., and Alvarez-Vincent, J.J. 1998. Synchronous glottic granular cell tumor and subglottic spindle cell carcinoma. Arch Otolaryngol Head Neck Surg. 124: 1031–1034

Le, B.H., Boyer, P.J., Lewis, J.E. and Kapadia, S.B. 2004. Granular cell tumor: immunohistochemical assessment of inhibin-alpha, protein gene product 9.5, S100 protein, CD68, and Ki-67 proliferative index with clinical correlation. Arch Pathol. Lab Med.128: 771-775.

Mazur, M.T., Shultz, J.J., and Myers, J.L. 1990. Granular cell tumor. Immunohistochemical analysis of 21 benign tumors and one malignant tumor. Arch Pathol. Lab Med.114: 692

Nagaraj, P.B., Ongole, R., and Bhujanga-Rao, B.R. 2006. Granular cell tumor of the tongue in a 6-year-old girl--a case report. Med Oral Patol Oral Cir Bucal. 11: E162-1624.

Neville, B.W., Damm, D.D., Allen, C.M., and Bouquot, J.E. 2002. Oral & Maxillofacial Pathology. 2nd ed. W.B. Saunders Co, Philadelphia. 12: 465-467.

Regezi, J.A., Sciubba, J., Richard, J., and Jordan, C.K. 2003. Oral Pathology: Clinical Pathologic Correlations. 4th ed. elsevier science. Saunders.

Roberto, A., Rejas, G., Campos, M., Cortes, A., Pinto, D., and De Sousa, S. 2011 Jan. The neural histogenetic origin of the oral granular cell tumor: An immunohistochemical evidence. Med Oral Patol Oral Cir Bucal. 1;16(1):e6-10. Oral granular cell tumor.

Silva, G.C., Vieira, T.C., and Vieira, J.C. 2007. Congenital granular cell tumor (congenital epulis): a lesion of multidisciplinary interest. Med Oral Patol Oral Cir Bucal. 12: E428-430.

Sposto, M.R., Navarro, C.M., and De Andrade, C.R. 2006. Granular cell tumour (Abrikossof tumour): case report. Oral. Oncol. Extra. 42: 194–197.

Vered, M., Carpenter, W.M., and Buchner, A. 2009. Granular cell tumor of the oral cavity: updated immunohistochemical profile. J. Oral Pathol Med; 38:150-159.

# ورم خلايا محببة باللسان
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**الملخص:**

ورم الخلايا المحببة هو ورم يصيب الأنسجة اللينة، وهو يعرف ويميز عن طريق الخلايا المكونة له والتي تتميز بوفرة السيتوبلازم المحبب.

وتؤكد الأبحاث الحديثة أن منشأ هذا الورم يأتي من الخلايا العصبية ولكن محفزات نشأته تبقى مجهولة.

ويصيب ورم الخلايا المحببة بشكل شبه حصري الفئة العمرية بين العقد الرابع والسادس، وغالبية المصابين به هم من النساء. ويعد ظهوره في الأطفال أمرًا غير شائع. وهذه الحالة نستعرض فيها ورم خلايا محببة في طفل سعودي ذكر يبلغ من العمر ١٣ سنة فقط، من الناحية الإكلينيكية والمخبرية، كما نقوم بمراجعة بعض النشرات العلمية بخصوص ورم الخلايا المحببة.

**الكلمات المفتاحية:** إصابة الأطفال، منشأ الورم من الخلايا العصبية، ورم خلايا محببة.