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 4th-6th 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: Childhood 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 4th to 6th 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.
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:


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الملخص:
ورم الخلايا المحبيبة هو ورم يصيب الأنسجة اللينة، وهو يعرف ويفتري عن طريق الخلايا المكونة له والتي تتميز بوفرة السيتوبلزم المحبيب.
وتؤكد الأبحاث الحديثة أن منشأ هذا الورم يأتي من الخلايا العصبية ولكن محفزات نشأته تبقى مجهولة.
ويصيب ورم الخلايا المحبيبة بشكل شبه حصري الفئة العمرية بين العقد الرابع والسادس، وغالبية المصابين به هم من النساء. وبعد ظهوره في الأطفال أمرًا غير شائع، وهذه الحالة تستعرض فيها ورم خلايا محبيبة في طفل سعودي ذكر يبلغ من العمر 13 سنة فقط، من الناحية الإكلينيكية والمخبرية، كما نقوم بمراجعة بعض النشرات العلمية بخصوص ورم الخلايا المحبيبة.

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