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Granular Cell Tumor of Tongue Base: A Rare Cause of Lump Sensation

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Abstract

Granular cell tumor is a rare neurogenic tumor originated oral cavity. Tongue is a main structure of the oral cavity. Because of various problems of tongue, chief complaints such as lump sensation and throat discomfort are presented. The symptoms affected granular cell tumors are non-specific. To remove this tumor and recover the quality of patient's life, a complete surgical excision is essential. It is important to differentiate granular cell tumor with other diseases to do appropriate treatment exactly. This study reports a successful treatment case of granular cell tumor of tongue base.

Keywords: Granular cell tumor; Neurogenic tumor; Tongue; Surgical excision

Introduction

Granular Cell Tumor (GCT), formerly called Abrikossoff's tumor, is considered as a benign tumor of the subcutaneous and submucosa area. Preferential localization is the head and neck mucosa, and especially the dorsal tongue mucosa [1]. This tumor is originated by nerve sheath tissue. The clinical expression of GCT is similar to other solitary, small and benign tumors of the oral mucosa. These tumors are painless and grow slowly, except rare syndromic forms [1,2]. Diagnosis is based on the pathological examination and immunohistochemical confirmation of the neural origin with S100 and CD68 expression [2]. Classical treatment for a GCT is a surgical excision with safety margins. Recurrence most often results from an incomplete resection or malignant forms. Benign form of GCT has an excellent prognosis, even though its pathogenesis remains uncertain. Granualr cell tumor should be distinguished from other diseases for any isolated nodule of the oral mucosa, particularly on the tongue. This study reported a successful treatment case of granular cell tumor of tongue base.

Patient Information

A 76-year-old male visited the Outpatient Clinic Department (OPD) complaining of lump sensation. The patient had no underlying systemic disease, previous surgical history, and masticating trauma history. The author checked him with physical and endoscopic evaluations. There was a 2 cm sized creamy lesion which is presented oval-shaped and painless mass on the left lateral border of tongue base **(Figure 1)**. Characteristic of the mass was single isolated, well-limited, and submucosal mass. The surface of mucosa is normal healthy state with grainy appearance and a lack of taste bud. There were not palpable lymph nodes of cervico-facial region at the physical evaluation. In consultation with the patient, a surgical mass excision was done. During the surgery, the surgical margin was secured over the borderline of mass and there was not a skipped mass lesion in the oral cavity. Pathologic finding was the focal neuromatous nerve tissue proliferation. After the surgery, the author evaluated him at an outpatient clinic. There was no recurrence of mass and the mucosal site of surgical lesion was well-healed. The chief complaint symptom of the patient was disappeared.

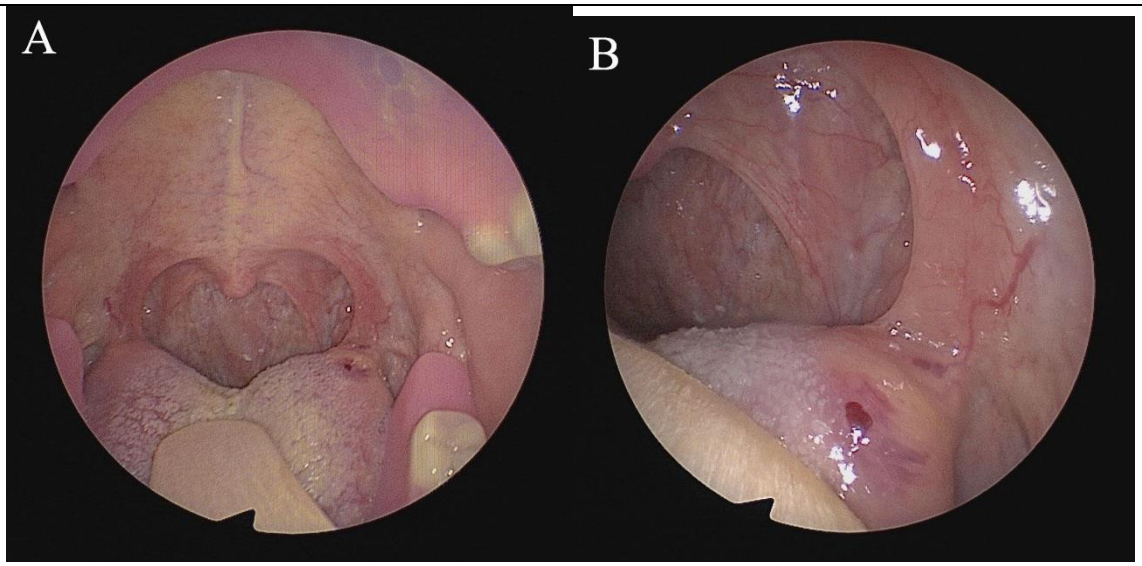

Figure 1: Endoscopic finding of neurogenic tumor. Neurogenic tumor lies on the left side of tongue posterior portion (picture A and B).

Discussion

GCT is a rare, benign soft tissue tumor, it is classified by WHO **[2]** among soft tissue and neural tumors in the section tumors of the oral cavity and mobile tongue. It was described for the first time by Abrikossoff in 1926 as a tumor of stratified muscle origin. It was initially called myoblastoma **[3]** because of the unencapsulated tumor and intermingling of the skeletal muscle bundles. Since then, it has acquired other names, including granular cell neurofibroma, granular cell schwannoma and Abrikossoff's tumor. Today, the immunohistochemical studies are consistent with Schwannian origin, which suggests a peripheral nerve sheath differentiation and neural origin **[2,4].** This hypothesis is in agreement with the ultrastructural

observation in electron microscopy studies [5,6] and the absence of the myoglobin in the granular cytoplasm of the cells. The etiopathogenesis of GCT remains uncertain, and some authors suggest a reactive origin [1]. GCT can affect any subcutaneous and submucosal sites in the body. The intraoral GCT are generally diagnosed between the 40 and 60 years old, with a peak in patients who are in their 50s [1,2]. However, rare pediatric cases have been reported [7]. There have been no reported cases in elderly subjects. Until previous reports, the oldest represented patient was 68 years old [8], but my data reported 76 years old patient case. Black populations are more affected than white population, and women are more affected than men, with female-to-male ration of 2:1 [2]. However, women represent more than 75% of GCT cases in this intraoral location [1]. The oral cavity is the most prevalent region of GCT in the literature. Of the oral cavity structures, tongue is the most common intraoral site of GCT occurrence, with more than 85% [1,2]. GCT usually takes place on the lateral border of tongue dorsum and less frequently arises on the tip of the tongue. The size of GCT is diverse, owing to slow-growing time. Actually, the average size is 1 cm. But there was a 3cm sized GCT in the reported paper.

Clinically, GCT of the oral mucosa appears as a single isolated lesion. However, it can also occur as multiple lesions [9], especially in the context of syndromes such Leopard syndrome, neurofibromatosis, Noonan syndrome, and Watson syndrome [10]. With the oral cavity, GCT was discovered incidentally because it grew up slowly and was asymptomatic. Otherwise, patients often express the abnormal symptoms such as throat discomfort and lump sensation. In the typical case of detected GCT, it presented well-limited, round or oval shaped submucosal mass. The surface mucosa is usually healthy with a slightly modified, grainy appearance and a lack of taste buds and the GCT is usually painless, firm, rubbery, well-limited, and non encapsulated in the physical evaluation [11] Occasionally, GCT mucosa can be creamy or yellowish colors. It can be caused by consumption of alcohol, tobacco, or nicorandil [12,13]. The patient of this case reported the symptoms such as lump sensation and throat discomfort when drinking and speaking. The size of the lesion remained small, about 2cm during the follow up period. The mucosal surface GCT lesion was healthy state. Because of the clinical symptoms of GCT is non-specific or asymptomatic, the differential diagnosis with GCT and other diseases is essential. It is the attentive point that there are the differential diseases, for example, neurofibroma, schwannoma, lipoma, or even a benign fibroepithelial hyperplasia [13]. Actually, because GCT is a benign tumor, radiological evaluation cannot be needed to. However, because occasional malignant transformation of GCT can be arisen rarely, radiological examination is essential to confirm the invasion adjacent to GCT and metastasis. Histopathological examination is important to do exact diagnosis and appropriate treatment **(Figure 2)**. GCT is an unencapsulated tumor whose cells intermingle with adjacent normal tissue by bands of connective tissue [11]. GCT is characterized by the presence of sheets and cords of large, polygonal, round, or elongated cells with indistinct membranes containing abundant eosinophilic granular cytoplasm which are PAS positive [11]. Also, there are architectural growths patterns can be distinguished in the GCTs. One pattern is a nodular type surrounded by the residual structure. In the nodular center, a large nerve trunk can be found. Another pattern features

cords and small nests infiltrating the surrounding mesenchymal structures, where only small nerve bundles are present. Both patterns are present in GCT **[15].** All these microscopical criteria are characteristic of the majority of typical GCTs, which are benign tumors with no metastatic potential, as reported by Fanburg-Smith et al **[16].** Also, there are several characteristics presented with atypical forms of GCTs, for example, mitotic figures, spindle cell formation, slight nuclear polymorphism and increased nuclear cytoplasmic ratio. These slight modifications remain far outside the spectrum of cytonuclear modifications of malignant forms, which present numerous unfavorable histological criteria, such as strong cellularity, cellular atypia, spiny cells, cellular polymorphism, an increased nuclear-cytoplasmic ratio with prominent nucleoli, and the presence of focal or extensive tumor necrosis **[17].** Nasser et al. proposed using the Ki-67 and p53 immunolabelling index to differentiate between benign and aggressive/malignant behavior. A rate of Ki-67 of 10% was suggested for a cutoff **[18].** But, Berge et al. reported that the Ki-67 index was less than 5%. Metastases are the only definitive criterion for malignancy in GCT regardless of the histopathologic characteristics **[11].** In the reported literatures, there are malignancy cases which are about 2% of cases of GCTs, rarely **[17].** Malignant transformation of GCT can induce from pseudoepitheliomatous hyperplasia with acanthosis on the overlying epithelium to well-differentiated cell carcinoma. According to published reports, there is no case of malignant transformation of GCT in the oral cavity. Some authors have reported that histopathological examination exhibits pathognomonic features, making an immunohistochemical examination unnecessary **[19].** However, Immunohistochemical examination have shown that granular cell has high reactivity for S100 protein, CD68, vimentin, and neuron-specific enolase (NSE). Immunohistochemical examination is essential to establish a definitive diagnosis because it is difficult to clinically diagnose GCT **[20].** Treatment of oral GCT is the complete surgical excision over the negative margins.

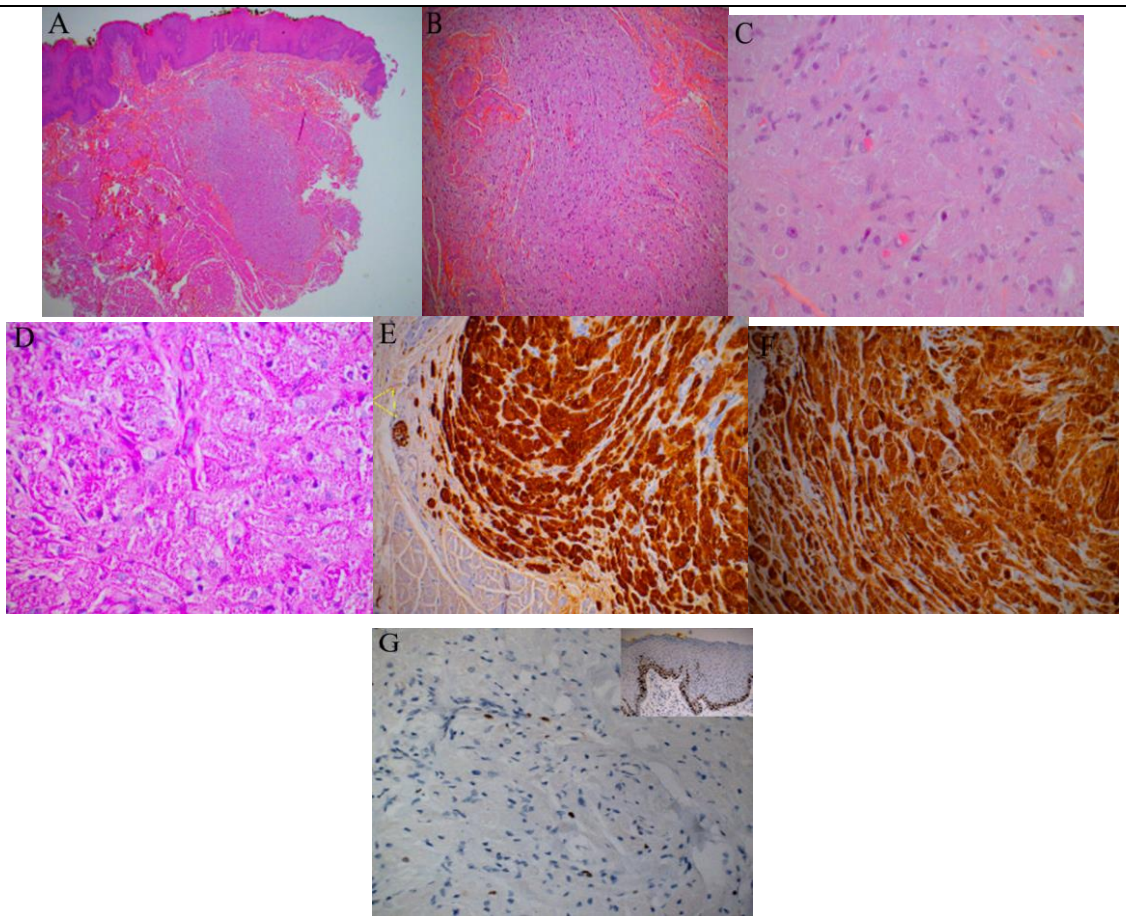

Figure 2: Representative histological and immunohistochemical findings. A and B. Hematein-Eosin-Saffron Stain revealed acanthosic epithelium and massive tumor well limited and no encapsulated with pseudo infiltrative and fibrosis aspect dissociating muscular bundles in the under the mucosa part and pushing effect in the deep part. (A: x2.5; B: x10 magnification). C. large granular cells with central small dark nuclei with an abundant eosinophilic cytoplasm. D. Periodic Acid-Schiff (PAS) stain revealed PAS-positive cytoplasmic granulation. E. strongly positive immunostaining for S100 (yellow head of arrow shown nervous thread as internal positive control) (x20 magnification) F. strongly positive immunostaining for CD68. G: Ki-67 immunolabelling shown absence of proliferation of tumor cell (mini-square: positive control in oral epithelium).The serial pictures are referenced by Berge C et al.

Conclusion

GCT is a benign neurogenic tumor originated oral cavity rarely. This disease usually has non-specific clinical symptoms. GCT presents well-limited, round or oval shaped submucosal mass in the oral cavity. GCT is mis-recognized in the normal healthy mucosa because of slow-growing, painless characteristics. GCT can be transformed in the malignancy, rarely. Therefore, it is important to do the differential diagnosis with other diseases and gold standard treatment of GCT is the complete mass excision.

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