

GRANULAR CELL TUMOR OF THE TONGUE: A RARE CASE REPORT

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ABSTRACT: Granular cell tumor (GCT) is a rare benign neoplasm that can appear in any site of the body, but most are located intraorally especially the tongue. It is a tumor of uncertain origin. Literature suggests that GCT has been associated to skeletal muscle, histiocytes, fibroblasts, myoepithelium, and nerve cell connective tissue origin. Today, following the introduction of immunohistochemistry, the hypothesis of neural origin has been more widely accepted. The GCT typically presents as a small solitary, slow growing, sessile, asymptomatic firm mucosal nodule with a smooth surface. Although majority of GCTs are benign, malignant form has been reported in 2% of cases. Even though it is a rare lesion, it must be included in differential diagnosis of oral lesions which are clinically similar to GCT for timely diagnosis and treatment. We report a rare case of GCT of the tongue in a 41 year old female with the emphasis on importance of a multidisciplinary approach to diagnosis and treatment.

KEYWORDS: Abrikossoff tumor, Intraoral Neoplasm.

INTRODUCTION

Granular cell tumor (GCT) is a rare benign soft tissue tumor that was first described in a patient with a lesion on the tongue by Abrikossoff (1926).¹ GCT can occur in almost any part of the body, with about 45–65% cases occurring in the head and neck region² and about 23–28% of head and neck GCTs are found involving the tongue.³ Literature suggests that GCTs has been associated to skeletal muscle, histiocytes, fibroblasts, myoepithelium, and nerve cell connective tissue origin.⁴ However it is now believed to be of primitive neuroectodermal origin.⁵ Since GCTs are small, solitary, slow growing painless tumor it often gets unnoticed and the diagnosis is often delayed. Hence it is important to consider GCTs under differential diagnosis of similar oral lesions for timely diagnosis and management. A rare case of GCT of the tongue in a 41 year old female with the emphasis on importance of a multidisciplinary approach to diagnosis and treatment is reported in this article.

Case report

A 41 years old female patient had presented with a history of a painless swelling in the right lateral margin of the tongue since 3 months. Intraoral examination revealed a solitary, smooth, sessile nodule about 2 cm. in size with a yellowish white cast on the right lateral margin of the tongue in relation to right lower molar teeth (**Fig.1**). The nodule was non tender and firm on palpation. Sharp edges of the lingual cusp of the molar teeth on the right side was noticed. A differential diagnosis of a traumatic fibroma of

the tongue, lipoma, oral lymphoepithelial cyst, pleomorphic adenoma of the minor salivary glands of the tongue was given. Selective grinding of the sharp edges of teeth was advised. Excisional biopsy was performed and histopathological feature (**Fig.2**) was consistent with a granular cell tumor. The postoperative course was uneventful. The patient was advised for review at six-month intervals to evaluate for recurrence and any malignant transformation.

Discussion

Abrikossoff in 1926 first named GCT as granular cell myoblastoma⁶ then later, it was termed as granulocellular rhabdomyoma by Cardenal and Oller (1947)⁷, as the tumor was thought to be of muscular origin.⁸ The histiocytic origin of GCT was proposed by Whitten⁸. Other investigators⁹ have proposed a neurogenic origin on the basis of the close association of the tumor with the nerves and ultrastructural findings of neurofilaments in the granular tumor cells. This theory was supported by Holland et al,⁸ who demonstrated S-100 staining in Schwann cells but not in myofibers. Today, on the basis of immunohistochemical studies, the hypothesis of neural origin has been more widely accepted.¹⁰ Thus, the GCT has received different names and has also been termed, tumor of Abrikossoff, myoblastoma, granular cell neurofibroma or granular cell schwannoma.⁵



Fig.1. Clinical appearance of granular cell tumor of the tongue.

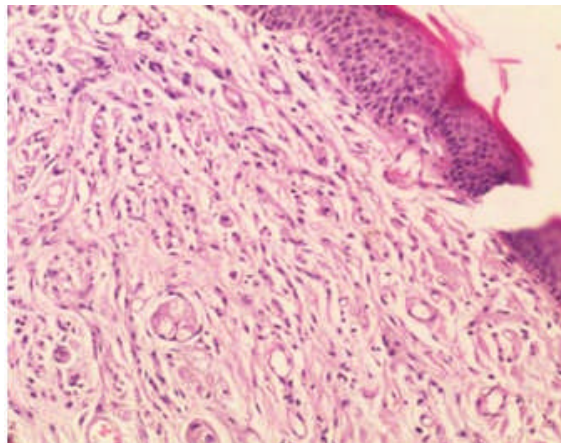


Fig.2. Photomicrograph showing typical histopathological aspect of the granular cell tumor, characterized by the presence of apparently normal pseudoepitheliomatous hyperplasia of the epithelium. The connective tissue stroma shows fibrillar connective tissue stroma with small islands and cords of large granular cells. (H&E stain 10X).

GCT is a rare benign neoplasm that occurs in different parts of the body such as the skin, nervous system, gastrointestinal tract, urinary bladder, female reproductive tract and bronchus.² The head and neck region are involved in about 45 to 65% of all GCTs with 70% presenting as intra oral lesions.¹¹ In addition to the oral mucosa, other sites in the head and neck region include the orbit, larynx and parotid gland.¹ Intraorally, the tongue, buccal mucosa, hard palate is commonly affected. The tumors of the lip, gingiva and uvula² have also been reported. On the tongue, the granular cell tumour usually occurs on the postero-lateral aspect.¹

Women are twice as commonly affected as men.² and more than half of the cases have occurred in African American patients.⁸ The age range varies widely from 4 months to 89 years, with a mean age between the fourth and sixth decades of life.⁸ Two distinct subtypes of GCTs are: 1) a congenital epulis, or a gingival GCT of infancy, and 2) a more common adult GCT.⁸

The GCTs typically presents as a solitary, slow growing, sessile, asymptomatic firm mucosal nodule of less than 3cm in size with a smooth surface. The surface color of the mucosal nodule varies from normal or slightly pale to yellowish.⁸ The nodule generally reveals an intact overlying epithelium. Large lesions may show surface ulceration which may clinically mimic a malignant lesion.² GCTs usually presents as a single lesion, but multifocal tumors at the first presentation are reported in 4–10% of cases.⁸

Histologically, granular cell tumor is poorly circumscribed and exhibit numerous strands and sheets of round / polygonal cells with distinct cellular membrane but cells may also be found arranged in cords and nests. The individual cells have abundant pale cytoplasm with eccentrically placed small nuclei and abundant eosinophilic coarse granules.¹ In lesions involving tongue, the pseudoepitheliomatous hyperplasia may be so pronounced that it has been misinterpreted as squamous cell carcinoma.¹²

The immunohistochemical studies of the GCT shows a strong and uniform positivity for S-100 protein,¹² a widely used marker for identification of the granular cells in GCTs. CD-68 is another traditionally used marker for identifications of GCT (2,19 in 1). Granular tumor cells can also express neurone-specific enolase, calretinin, inhibin-alpha and PGP 9.5, but no reactivity for smooth muscle actin.¹²

Clinically the appearance of GCT is indistinguishable from other benign connective tissue tumors such as fibromas, lipomas, neuromas, neurofibromas or schwannomas with their malignant variants, minor salivary gland tumors.¹ Other conditions that could be included are oral carcinomas, amyloidosis, colloid milium, sarcoidosis, aspergillosis, and Urbach-Wiethe disease.¹² Majority of the GCTs are benign, but approximately 2% can present with malignant behaviour.² A GCT is considered to present with uncertain malignant potential if the clinical size of tumor is over 4cm as is the ability to infiltrate underlying and adjacent structures, high speed of growth, presence of regional and distant metastasis and rapid recurrence and

histologic data of malignancy.⁷ The imaging studies, CT scan and/or MRI, may detect occult metastases.⁷ The most common site of metastasis are lymph nodes, bony tissue, peripheral nerves, peritoneal cavity, and lungs.¹² In the present clinical case we present, there are no clear histological parameters of malignancy.

The treatment of choice is complete surgical excision with a margin of safety and the prognosis is favorable due to its slow growth rate, low aggressiveness and low recurrence rates.^{5,8} In the case of malignant GCT, the recurrence rate is high (70 percent).¹² Although complete surgical excision leads to cure, a careful follow up is necessary in order to diagnose possible recurrences (1-3% of cases) and to assess for malignant trans-formation.

CONCLUSION

An uncommon case of GCT involving the tongue is reported in a 41year old female patient. Even though it is a rare lesion, a clinician must know about its occurrence and it must be included as a part of differential diagnosis of oral lesions which are clinically similar to GCT. Since mass formation is the only subjective symptom and no characteristic clinical signs are induced by GCT, it is diagnosed on the basis of histopathological findings. Furthermore, immunohistochemical assay can make the diagnosis more precise. The patient should be reassured of the benign nature of the tumor and a careful follow-up is necessary in order to diagnose possible relapses.

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