CASE REPORT

Granular cell tumor of the tongue: Report of a case

Alka Dive, Akshay Dhobley, Prajakta Zade Fande, Sudhanshu Dixit[1]
Department of Oral and Maxillofacial Pathology, VSPM's Dental College and Research Centre, Nagpur, [1]Oral and Maxillofacial Pathology, Rungta College of Dental Science and Research Centre, Bhilai, Chhattisgarh, India

Address for correspondence:
Dr. Akshay Dhobley,
Department of Oral and Maxillofacial Pathology,
VSPM's Dental College and Research Centre,
Nagpur, Maharashtra, India.
E-mail: drakshay94@gmail.com

ABSTRACT

Granular cell tumor (GCT) is a benign lesion characterized by the accumulation of plump cells with abundant granular cytoplasm. The formation of a granular cell tumor is a neoplastic process and the lesions formed are of neural derivation, as supported by immunophenotypic and ultra structural evidence. This type of tumor has been found to be both benign and malignant although malignancy is rare and comprises only 2% of all granular cell tumors. Here we report a case of GCT in a 40 year old male patient on the posterolateral border of tongue.

Key words: **Tongue, granular cell tumor, pseudoepitheliomatous hyperplasia**

INTRODUCTION

In 1926 Arbikossoff described a tumor of the tongue composed of granular cells derived from striated muscles and termed it as granular cell myoblastoma, theory that was subsequently abandoned. Granular cell tumor (GCT) is a benign lesion characterized by the accumulation of plump cells with abundant granular cytoplasm. A wide variety of cell types have been proposed as the cells of origin, including histiocytes, fibroblasts, myoblasts, neural sheath cells, neuroendocrine cells, and undifferentiated mesenchymal cells.[1]

GCTs can affect any part of the body, however, in head and neck area it predominats by 45% to 65%. Of the head and neck cases, 70% of lesions are located intraorally (tongue, oral mucosa, hard palate). GCTs are typically small, solitary lesions; rarely do they exceed 3 cm in size. Both benign and malignant lesions have been reported; although malignancy occurring is rare, comprising of 2% of all GCTs.[2]

Here we report a case of GCT of tongue.

CASE REPORT

A 40-year-old male reported with a chief complaint of painless swelling on the left lateral border of tongue since one year. The swelling was initially small in size and slowly progressed to present size of 1 × 1 cm. The patient gave no history of trauma, pain, bleeding, ulceration or pus discharge. Past dental history, family history and drug history was not contributory. Patient gave history of tobacco chewing and bidi smoking (4-5/day) since 15 years.

Intraoral examination revealed a well circumscribed soft tissue swelling with a smooth surface, oval shape and measuring 1 × 1 cm approximately on the left posterolateral aspect of lingual dorsum. The swelling was pinkish pale white in color. Characteristic loss of the gustatory papillae of the overlying mucosa was seen.

On palpation, swelling was firm, mildly tender, adherent to the underlying structures with no regional lymphadenopathy.

A clinical diagnosis of Fibroma was made. Routine hematological investigations revealed normal values. Excisional biopsy was performed and specimen was sent for histopathological examination.

Gross examination revealed a soft tissue mass, pale creamish to brownish in color, with well defined borders, 1 × 1 cms, oval in shape and soft in consistency [Figure 1].

Histopathological examination of H and E stained section revealed lesional tissue consisting of parakeratinized stratified squamous epithelium showing pseudoepitheliomatous hyperplasia The underlying connective tissue shows tumor cells arranged in the form of nests and ribbons [Figures 2 and 3]. These neoplastic cells are large polygonal in shape with indistinct cell borders, abundant coarse eosinophilic granular cytoplasm and pale eccentric nuclei, some of which are vesicular. These neoplastic cells exhibit mild hyperchromatism

Access this article online

Quick Response Code:

Website: www.jomfp.in

DOI: 10.4103/0973-029X.110728

and pleomorphism [Figure 4]. The adjacent fibro-cellular stroma consists of collagen fibers, fibroblasts and few endothelium lined blood capillaries.

Based on histopathological findings the final diagnosis of GCT was made.

DISCUSSION

Granular cell tumor (GCT) is a benign lesion that occurs at various sites of the body with preponderance to the oral cavity. The histogenesis of GCTs has remained enigmatic in spite of a vast number of immunohistochemical and ultra structural studies. A neural origin or differentiation, in particular of the schwann cell type, is currently in favor. This is based on the close anatomical relationship of GCTs to peripheral nerve fibers; on the ultrastructural demonstration of myelinfigures; axon-like structures and on its immunohistochemical reactivity with S-100 protein, neuron-specific enolase and myelin proteins.[1] The tongue is rich in nerve fibers encased by schwann cells that can be the source for GCTs. GCT cannot be morphologically distinguished from tumors of muscle cell origin following an event of metabolic distress.[3] GCTs from various sites of the body were recently found to be positive to a number of new markers of alleged neural differentiation as well as to several non-neural markers, such as inhibin-α.

Oral GCTs shows a female predilection (F:M = 2:1) and the age range is from childhood to elderly, with a mean age of occurrence in the fourth decade of life and 67-81% of the lesions occurr in tongue.[1,3] In agreement, our case of GCT was seen in a patient who was 40 years old with the presentation site being the posterolateral aspect of tongue; however, the patient was male.

Benign GCTs are generally solitary asymptomatic nodules, pink in color occasionally yellowish, less than 3 cms involving the subcutaneous or submucosal tissues. The nodular mass is hard and reveals an intact overlying epithelium.[2] In our case characteristic loss of the gustatory papillae and depapillation of dorsum of tongue was seen.

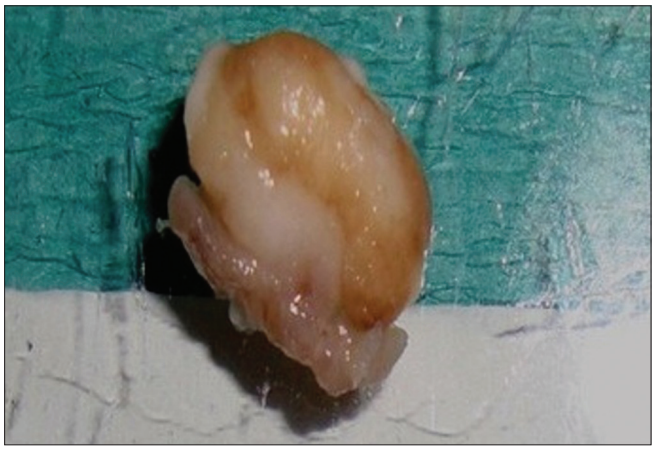

Figure 1: Gross examination revealed a soft tissue mass, pale creamish to brownish in colour, with well defined borders, 1 × 1 cms, oval in shape and soft in consistency

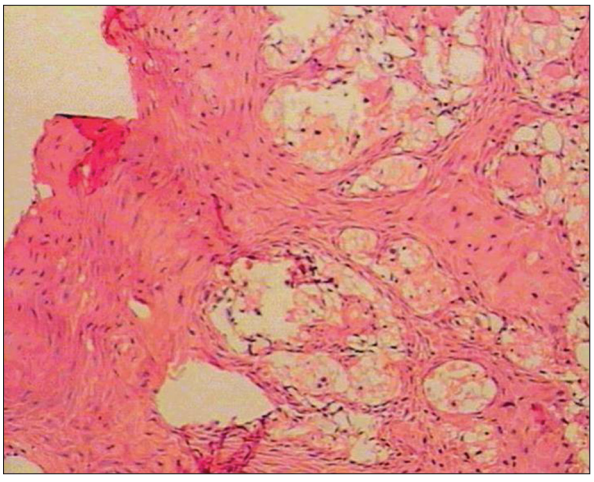

Figure 3: Photomicrograph showing parakeratinized stratified squamous epithelium with pseudoepitheliomatous hyperplasia. (H and E, ×100)

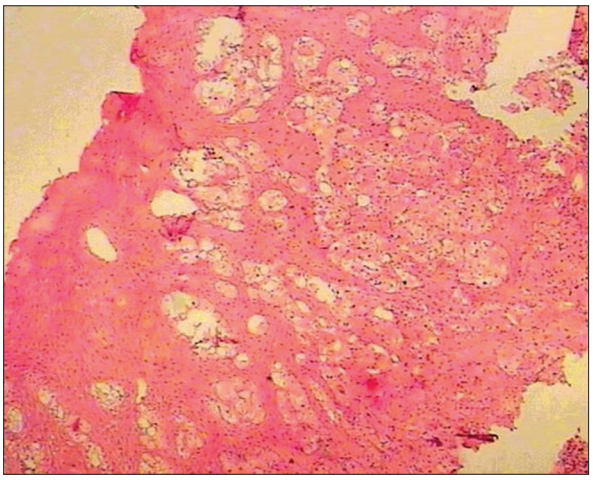

Figure 2: Photomicrograph showing lesional tissue consisting of epithelium and connective tissue. (H and E, ×40)

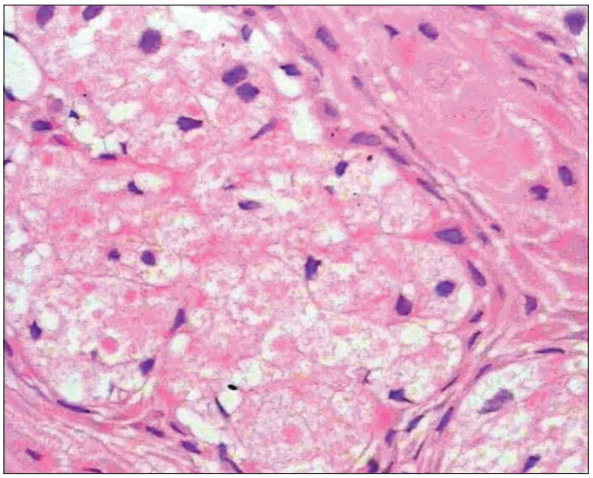

Figure 4: Photomicrograph showing polygonal neoplastic cells exhibiting mild hyperchromatism and mild pleomorphism with indistinct cell borders, containing pale eosinophilic granular cytoplasm and round to oval, eccentrically placed nucleus. (H and E, ×400)

Morphological assessment

The following morphological parameters were assessed on the haematoxylin and eosin-stained slides.

Architecture of the lining of the oral epithelium – exhibiting acanthosis and pseudoepitheliomatous hyperplasia (PSE), florid PSE.
Lesion's relationship to the lining of the oral epithelium – The lesional cells were in close relation to the oral epithelium.
Lesion's architectural pattern – circumscribed nodule lacking infiltrative characteristics.
Surgical margin status – Was free of tumor.

Oral GCTs exhibit pseudoepitheliomatous hyperplasia (PSE) in about 50% of the cases. It has been suggested that stimulation of basal cell proliferation occurs through an interaction between the granular cells and the neighbouring epithelial cells.[1] In our case, marked PSE was present mimicking squamous cell carcinoma (SCC). However, SCC was ruled out from diagnosis due to absence of severe malignant features like nuclear pleomorphism, increased nuclear cytoplasmic ratio, mitotic figures, hyperchromatism, etc., The pathologist must be very cautious about misdiagnosing this as well-differentiated SCC and, by definition a carcinoma-like surface lesion with underlying granular cells should be considered to be a benign, reactive change. No granular cell tumor of the mouth has yet been associated with a true squamous cancer and the lingual dorsum is one of the oral sites least likely to develop such a cancer.

Campbell (1955) stated that the granular cell tumor and congenital epulis are practically identical. The pseudo-epitheliomatous hyperplasia is of course characteristic of the GCTs, but it is noteworthy that at no stage is a transition observed between such epithelial cells and the underlying granular cells. Apart from the presence of the granular cells in the congenital epulis, one of the most striking features is the presence of atrophic epithelium and high degree of vascularity to the stroma,[4] which was not present in our case therefore congenital epulis was ruled out from the diagnosis.

Reports on the histomorphologic features of GCTs are inconsistent. Some authors, report that nuclei of GCTs are dark and hyperchromatic, while others report that they are pale and vesicular. Some authors claim that the nuclei are located in a central position, while others describe them in an eccentric position.[1] This case revealed that granular cells were round, oval or polygonal with both vesicular and eccentrically placed nuclei.

Special staining

P.A.S. Using the technique of Hotchkiss weakly positive results were obtained. The color obtained within the granules by this method was not altered by application of diastase.

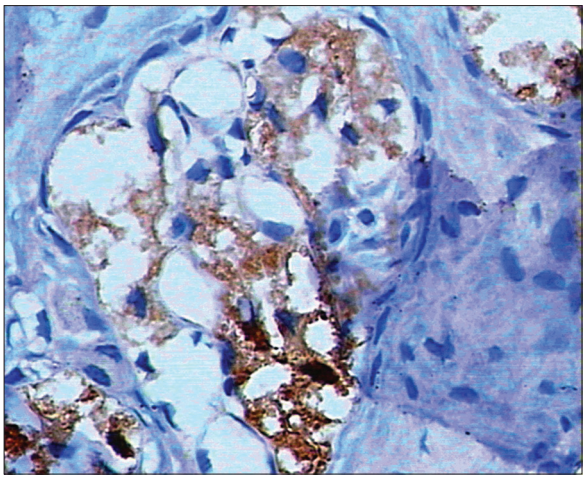

Figure 5: Photomicrograph showing strong positivity of granules for the S-100 protein (S100, ×400)

Immunohistochemistry

The lesion was strongly positive for the S-100 protein, which can be used to serve as sound evidence for their neural origin [Figure 5].

The malignant GCTs have two distinct variations. The first variant has a benign histopathology, except for increased mitotic activity and mild nuclear pleomorphism. The clinical features of large size, rapid growth and surface ulceration must therefore, be used to arrive at a malignant diagnosis and the pathologist should carefully evaluate the lesional periphery for signs of true invasion. The second variant shows transition from typical benign granular cells to pleomorphic granular cells to pleomorphic non granular spindle cells and giant cells with numerous mitotic figures. Constellation of histologic features portends an increased risk for metastasis. Such features include necrosis, spindling, vesicular nuclei with prominent nucleoli, increased mitotic activity (>2 mitoses/10 HPF), high nucleo-cytoplasmic ratio and pleomorphism. Tumors with three or more of these features are considered malignant and have approximately 40% risk of causing death.[5]

Conservative excision is the treatment of choice for granular cell tumor. A few reported metastasizing GCTs have appeared to be histologically benign and for this reason, tumors which recur, grow rapidly or reach a size greater than 5 cm. should be viewed with grave suspicion.[5]

In summary, morphologically oral GCTs may demonstrate a wide variety of features and architectural patterns, but they all still exhibit a benign behaviour. Furthermore, GCTs could be regarded as lesions that reflect a local metabolic or reactive change rather than a true neoplasm.

REFERENCES

1. Vered M, Carpenter WM, Buchner A. Granular cell tumor of the oral cavity: Updated immunohistochemical profile. J Oral

Pathol Med 2009;38:150-9.
2. Nagaraj PB, Ongole R, Bhujanga-Rao BR. Granular cell tumor of the tongue in a 6-year-old girl- a case report. Med Oral Pathol Oral Cir Bucal 2006;11:E162-164.
3. Stewart CM, Watson RE, Eversole LR. Oral granular cell tumors: A clinicopathologic and immunocytochemical study. Oral Surg Oral Med Oral Pathol1988;65:427-435.
4. O'brien FV. Granular cell lesions of the mouth. Br J Oral Surg 1972;10:89-94.
5. Enzinger FM, Weiss SW. Soft Tissue Tumors, 4th ed. St Louis: Mosby; 2001. p. 1178-87.

How to cite this article: Dive A, Dhobley A, Fande PZ, Dixit S. Granular cell tumor of the tongue: Report of a case. J Oral Maxillofac Pathol 2013;17:148.

Source of Support: Nil. **Conflict of Interest:** None declared.