Case Report

Atypical Presentation of Granular Cell Tumor Involving Tongue: A Rare Case Report

Abstract

Granular cell tumor (GCT) is a rare benign tumor chiefly affecting the orofacial region, especially tongue. The origin of this neoplasia, after remaining a controversy for years, was finally identified to be from Schwann cell or neuroendocrine cell. They usually present as asymptomatic, firm, sessile, submucosal, solitary, or multiple nodule/papule like lesions. Histopathologically, the presence of sheets, cords or nests of large cells having abundant, eosinophilic granular cytoplasm, and pseudoepitheliomatous hyperplasia are the characteristic features of this neoplasm. Immunohistochemical stains such as S100, Cluster of differentiation 68, neuron-specific enolase, and laminin also aid in the proper diagnosis of granular cells. We hereby present a case of GCT in a 50-year-old male, which had a unique exophytic appearance, probably being only the second to be reported till date.

Keywords: *Exophytic, granular cell tumor, immunohistochemistry, tongue*

Sudeshna Bagchi, Sanchita Kundu, Arunit Chatterjee, Rudra Prasad Chatterjee

Department of Oral and Maxillofacial Pathology, Guru Nanak Institute of Dental Sciences and Research, Kolkata, West Bengal, India

Introduction

Granular cell tumor (GCT) or Abrikosoff's tumor is an uncommon benign soft-tissue neoplasm which commonly occurs in the head-and-neck region.[1,2]

Histogenesis of GCT is uncertain. Many cell types, including muscular, mesenchymal, Schwann cell, neuroendocrine (NE) cell, and histiocytes,[3,4] have been implicated regarding the origin of this neoplasm, and thus different nomenclatures have been applied to this rare entity such as myoblastoma, granular cell neurofibroma, and granular cell schwannoma by various authors.[3-8]

Overall, the incidence of this rare neoplasm is about 0.03%,[3] out of which 70% of the lesions are found intraorally[1] where the tongue is the most common site.[2] Other sites such as hard palate, buccal mucosa, lip, uvula, parotid gland, and gingiva have also been reported.[2] GCTs can also involve a wide variety of anatomical sites such as orbit, larynx, parotid gland, breast, gastrointestinal tract, reproductive systems, and nervous system.[5,6] It is commonly found in the 4th–6th decades of life[2,5,7] and females are affected more than males in a ratio of 2:1.[7] The lesion usually presents as asymptomatic, firm, sessile, submucosal, solitary or multiple nodules/papules, pink or yellowish in color, usually 2 cm or less in diameter and typically has a smooth surface.[4,5,7-9] In 2012, Cole *et al*. reported an atypical presentation of GCT which was located on the dorsum of the tongue and was firm, pedunculated, polypoid, and tender.[5] Clinically, GCTs should be differentiated from some other neoplasms such as fibromas, lipomas, neurofibromas, neuromas, or schwannomas.[3,5]

Histopathologically, GCTs are composed of large, polygonal, or fusiform cells with abundant, pale, eosinophilic, granular cytoplasm, and small vesicular nuclei arranged in sheets, cords, or nest-like fashion.[4,7,9] The granules are periodic acid–Schiff positive and diastase resistant.[9] Ultrastructural studies had depicted the cytoplasmic granules as autophagic vacuoles containing cellular debris, including mitochondria and fragmented endoplasmic reticulum along with myelin.[9] An unusual and significant microscopic finding (50%)[1,2,7] is the presence of pseudoepitheliomatous hyperplasia (PEH) along with acanthosis of the overlying epithelium which is sometimes misdiagnosed with squamous cell carcinoma.[1,2,9] Malignant transformation of this neoplasm is very rare, about 1%–

How to cite this article: Bagchi S, Kundu S, Chatterjee A, Chatterjee RP. Atypical presentation of granular cell tumor involving tongue: A rare case report. Int J App Basic Med Res 2021;11:111-3.

Submitted: 30-Dec-2019
Revised: 02-Dec-2020
Accepted: 31-Jan-2021
Published: 08-Apr-2021

Address for correspondence:
Dr. Sudeshna Bagchi, Department of Oral and Maxillofacial Pathology, Guru Nanak Institute of Dental Sciences and Research, 157/F Nilgunj Road, Panihati, Kolkata - 700 114, West Bengal, India.
E-mail: sudeshna.bagchi2@gmail.com

Access this article online

Website: www.ijabmr.org

DOI: 10.4103/ijabmr.IJABMR_438_19

Quick Response Code:

2%, which is characterized by nuclear pleomorphism and hyperchromatism along with necrosis and increased mitotic activity.[10]

Immunohistochemically, GCTs are S-100, neuron-specific enolas (NSE), laminin, myelin basic protein, and cluster of differentiation (CD)-68 positive; and negative for neurofilament protein and glial fibrillary acidic protein.[2-4,7,9]

Case Report

A 50-year-old male patient from semi-urban area reported to our department with a chief complaint of a swelling involving the tongue for the past 6–7 months. The swelling was small and asymptomatic initially and had gradually increased to the present size.

Intraoral examination revealed the presence of a solitary, sessile, exophytic, nodular growth involving the middle third of the dorsum of the tongue measuring 1 cm × 1 cm × 0.5 cm, covered by normal oral mucosa in association with an elliptical depapillated area [Figure 1a]. On palpation, the growth was nontendered, firm, nonpulsatile, noncompressible, and nonreducible in nature.

Based on the clinical presentation, a provisional diagnosis of fibroma was made. The differential diagnoses included papilloma, GCT, and neurofibroma.

The patient was advised to report with routine hematological and biochemical investigations. All parameters were within the normal limits. An excisional biopsy of the lesion was performed under local anesthesia, and the excised specimen was sent for the routine histopathological examination.

Sections stained with hematoxylin and eosin revealed the presence of hyperparakeratotic stratified squamous surface epithelium showing acanthosis and PEH, supported by underlying fibrovascular CT stroma. The most striking feature was the presence of large, polygonal cells in sheets with abundant, pale, eosinophilic, granular cytoplasm, dark vesicular nuclei, and indistinct cell borders in the CT. These cells were also found in association with the underlying muscle tissue [Figure 2a-c]. The overall clinicopathological features were suggestive of "GCT."

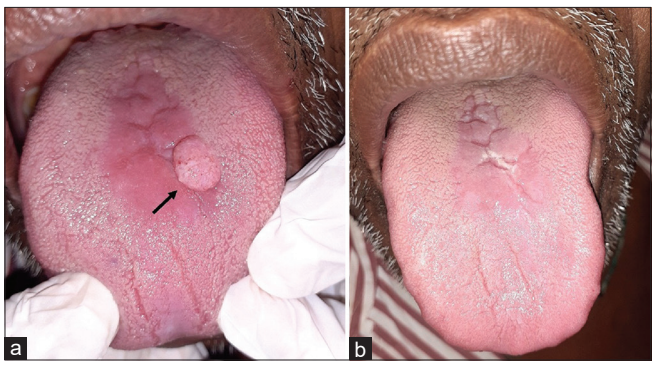

Figure 1: (a) Intra-oral photograph showing the presence of a solitary, sessile, exophytic nodular growth in the middle-third of the dorsum of the tongue (Black arrow) (b) Intra-oral photograph of the patient 6 months after the treatment

IHC evaluation was also done to confirm the diagnosis where the granular cells stained positively with S-100 [Figure 3a] and CD-68 [Figure 3b] markers, and the case was finally diagnosed with GCT.

The patient was on follow-up until complete healing was achieved. Postoperatively, no recurrence was reported up to 6 months [Figure 1b].

Discussion

GCT is a rare benign neoplasia of controversial origin. It was first described in 1926 by a Russian pathologist, Abrikossof, who named it "myloblastenmyome," a term reflecting skeletal origin. The apparent association also led to similar names such as granular cell myoblastoma and granulocellular rhabdomyoma.[6]

The origin of this lesion has remained a matter of debate for the decades. Apart from histogenesis from varying cell lines, several nonneoplastic theories such as inflammatory, degenerative, regenerative, and congenital causes were also suggested.[3] However, recent studies are mostly indicating toward a neural origin, chiefly from Schwann cell or NE cell.[1,7] This can be substantiated by the close anatomical relationship of GCT to peripheral nerve fibers, the ultrastructural demonstration of myelin figures and axon-like structures, immunohistochemical reactivity with S-100, NSE and myelin proteins[1,6] and positive staining with Luxol fast blue, a myelin stain.[6] Thus, a much generalized term "GCT" is used nowadays.[7]

The patient under discussion was a 50-year-old male having a solitary, sessile, exophytic growth on the dorsum of the tongue. As it affects females more, the occurrence of this lesion in a male patient was uncommon. A remarkable feature noted in the present case was the exophytic growth of the lesion, as the lesion is mostly submucosal as described by various authors.[1,2,8] Available literature indicates only one such reported case so far.[5] Other epidemiological and clinical features of the present case were all consistent with those mentioned in previous literatures.[4,5,7,8] Multiple or multifocal lesions can also be observed in 5%–16% of the cases.[10]

Light microscopic features of the present case revealed PEH and sheets of large, polygonal cells in the CT having abundant, eosinophilic, granular cytoplasm, and were in accordance with that described by authors of different studies.[1,2,4,7,9] Background stroma was minimal. With time, lesions become more desmoplastic along with scattered nests of granular cells in a dense fibrotic stroma.[9] The authors have suggested that the interaction between the granular cells and the neighboring epithelial cells stimulates the basal cell proliferation resulting in the development of PEH.[1,2]

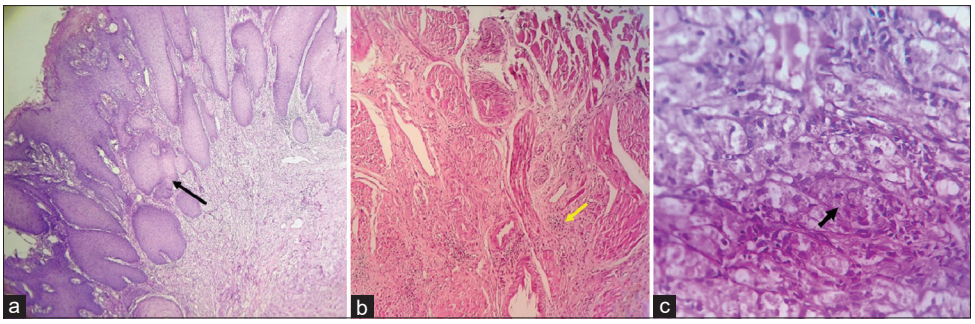

Figure 2: (a and b) Low-power photomicrographs (H and E, ×10) showing parakeratotic stratified squamous surface epithelium with pseudoepitheliomatous hyperplasia (black arrow) and acanthosis and sheets of large polygonal cells in the connective tissue in close association with the muscle layer (Yellow arrow) (c) High-power photomicrograph (H and E, ×40) showing large polygonal cells with abundant, pale, eosinophilic cytoplasm (black arrow)

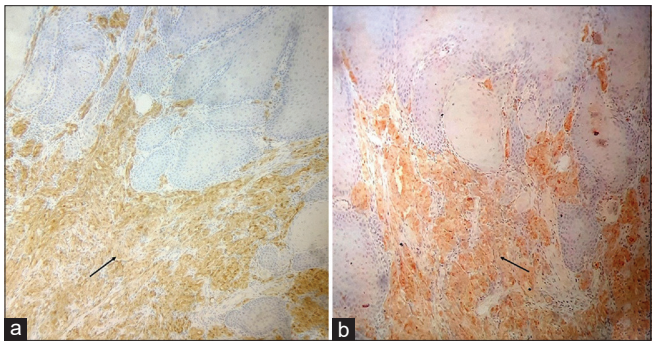

Figure 3: Low-power photomicrographs showing positive immunostaining of the granular cells with S100 (a) and CD68 (b) (black arrow)

Literature has described six histologic criteria to define malignant, atypical, and benign GCTs. These are necrosis, cell spindling, vesicular nuclei with large nucleoli, increased mitotic rate (>2/10 high-power fields), high nuclear-cytoplasmic ratio, and nuclear pleomorphism. The presence of three or more criteria defines malignancy, one or two defines atypia, and none or only pleomorphism defines benign lesion.[10]

Immunohistochemically, GCT shows positive immunostaining with S-100, CD-68, NSE, laminin, and myelin basic protein.[2-4,7,9] Our case also showed positivity with S-100 and CD-68. Positivity of GCT by vimentin, NK1/C3, Inhibitin-alpha, Calretinin, and protein gene product (PGP 9.5) has also been reported.[4] PGP 9.5 is presently being considered as the new marker of GCT.[4]

Conservative surgical excision is the treatment of choice of GCT. An excisional biopsy was performed in the present case which served as both diagnostic and remedial.

Thus, the clinical presentation, characteristic histological features accompanied by the positive immunohistochemical staining with S-100 and CD-68 confirmed the diagnosis of our case as GCT.

Recurrence is uncommon in GCT (7%),[9] which is frequently associated with incomplete surgical excision. Hence, proper care must be taken during surgical treatment, and long-term follow-up is essential to prevent malignant transformation and distant metastasis, which occur in 10% and 2%, respectively.[6]

Conclusion

GCT usually presents as sessile, submucosal, solitary/multiple nodular growth. However, in contrast to the usual clinical presentation, an exophytic nodular growth can also be seen, as in the present case, which stands unique. Hence, special attention should be given while diagnosing these lesions.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. 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.
2. Ferreira JC, Oton-Leite AF, Guidi R, Mendonça EF. Granular cell tumor mimicking a squamous cell carcinoma of the tongue: A case report. BMC Res Notes 2017;10:14.
3. Barbieri M, Musizzano Y, Boggio M, Carcuscia C. Granular cell tumour of the tongue in a 14-year-old boy: Case report. Acta Otorhinolaryngol Ital 2011;31:186-9.
4. Haikal F, Maceira J, Dias E, Ramos-E-Silva M. Histogenesis of Abrikossoff tumour of the oral cavity. Int J Dent Hyg 2010;8:53-62.
5. Cole E, Rahman N, Webb R. Case series: Two cases of an atypical presentation of oral granular cell tumour. Case Rep Med 2012;2012:159803.
6. Kumar S, Nair V, Ravindran R. A rare presentation of granular cell tumor of tongue – A case report. IOSR J Dent Med Sci 2017;16:102-7.
7. Neville BW, Damm DD, Allen CM, Chi A. Oral and Maxillofacial Pathology. 1st ed. South Asia, India: Elsevier; 2015. p. 502-3.
8. Eguia A, Uribarri A, Gay Escoda C, Crovetto MA, Martínez-Conde R, Aguirre JM. Granular cell tumor: Report of 8 intraoral cases. Med Oral Patol Oral Cir Bucal 2006;11:E425-8.
9. Shafer WG, Hine MK, Levy BM. Cyst and tumors of odontogenic origin. In: Rajendran R, editor. Shafers Textbook of Oral Pathology. 7th ed. New Delhi, India: Elsevier; 2012. p. 193-4.
10. Rose B, Tamvakopoulos GS, Yeung E, Pollock R, Skinner J, Briggs T, *et al*. Granular cell tumours: A rare entity in the musculoskeletal system. Sarcoma 2009;2009:765927.