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Case Report

Granular Cell Tumour of the Tongue: A Case Report of the Rare Lesion.

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Abstract

Granular Cell Tumour (GCT) is a rare and painless benign soft tissue neoplasm of Schwann cell derivative. It is a slow-growing, firm, and solitary nodule usually smaller than 3 cm in diameter that frequently occurs in females between 4^{th}-6^{th} decade of life. We aim to describe the clinical presentation of the granular cell tumour (GCT) and the histopathological tools used to achieve its definitive diagnosis.

We present a case of GCT of the tongue in a 46-year old female who presented with a year-old painless swelling on the tongue in our dental clinic. There was associated tenderness but disturbed deglutition. Clinical examination revealed a firm swelling in the right anterior two-thirds. Differential diagnoses made were chondroma, fibroma, and rhabdomyoma. Excisional biopsy for histopathological and immunohistochemical evaluation was diagnostic of GCT.

GCT may be confused with other oral lesions clinically, but the use of histopathological and immunohistochemical tools confirmed its diagnosis.

Keywords: Granular cell, immunohistochemistry, tongue, tumour,

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Introduction

Granular cell tumor (GCT) is a rare soft tissue tumour that arises throughout the body, most often in the oral cavity, gastrointestinal tract, subcutaneous tissue, and skin.[1,2] This tumor was first described in 1926 by the Russian pathologist Abrikossoff, hence the name Abrikossoff's tumor. Recent advances in electron microscopy and immunohistochemistry suggest that this tumor is derived from Schwann cells; thus, the name granular cell schwannoma.[1-4] It affects all age groups, with a female predominance in the 4th to 6th decades of life.[4] It represents 0.5% of all soft tissue tumours;[5] most often occurring as a solitary, painless nodule about 3-4 cm in diameter, and 7%-29% are multiple lesions.[2] The aetiopathogenesis is not fully understood, but recurrent genetic mutations have been reported in specific syndromes such as Noonan syndrome, Neurofibromatosis I, and LEOPARD syndrome.[2] Most of these tumors are benign, occurring more often among African Americans, while about 1%-2% are malignant with a poor prognosis, and 70% of the latter present in Caucasians. The head and neck are the most common sites of occurrence (45-60%), while the skin and subcutaneous tissue make up 30%.[2] We present a rare case of a lingual granular cell tumor in a 46-year-old female.

Case

A 46-year-old woman presented to our clinic on March 15, 2025, with a one-year history of a painless, slow-growing tongue swelling. There was no associated pain or tenderness, except for the bulge, which impaired deglutition. Clinical examination revealed an elevation in the middle third of the right half of the tongue. The overlying mucosa was normal, and the swelling was firm on palpation, measuring 3 cm in its widest diameter (Figure 1). Differential diagnoses included fibroma, chondroma, and rhabdomyoma. The patient underwent excisional biopsy under local anesthesia and was discharged after complete healing of the tongue (Figure 2).

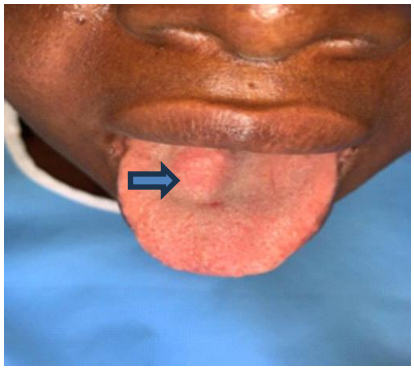


Figure 1: Right-sided tongue mass (blue arrow)

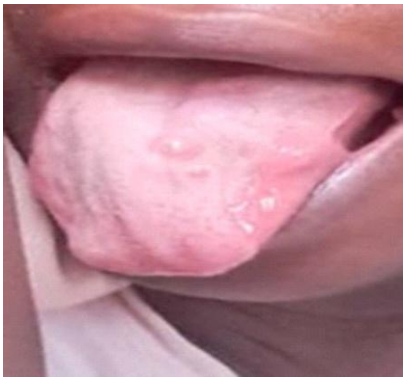
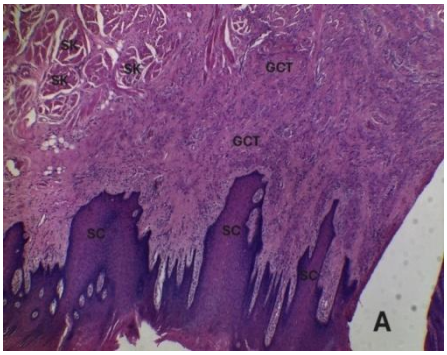


Figure 2: Healed site one-month post-excision

The specimen was submitted for histopathologic examination, and a definitive diagnosis of granular cell tumor was established on routine hematoxylin and eosin (H&E) stains (Figures 3 & 4).



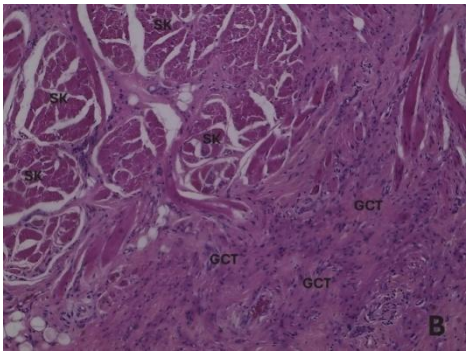
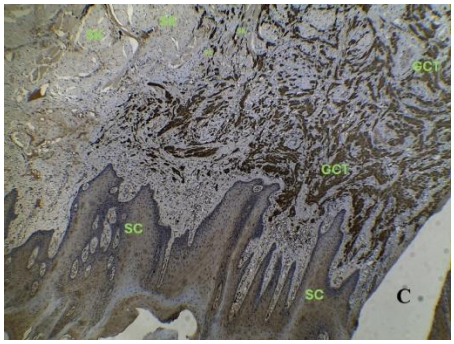


Figure 3 (A –H&E x4 obj) & figure 4 (B – H&E x10 obj): Photomicrographs showing syncytial proliferation of the granular cells (GCT) with pseudoepitheliomatous hyperplasia of the overlying squamous epithelium (SC), and adjacent normal skeletal muscle bundles (SK) of the tongue. The diagnosis was further assessed and confirmed by two immunohistochemical stains (IHC): S-100 (Figures 5) and CD68 (Figures 6).



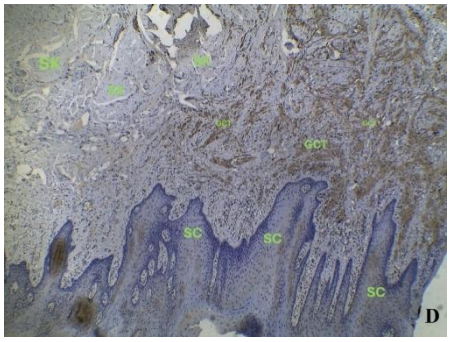


Figure 5 (C – S-100 IHC x4 obj) & figure 6 (D – CD68 x4 obj): Photomicrographs showing positive IHC cytoplasmic stain displaying deep brown pigmentation of the tumour cells (GCT).

Discussion

GCT is an uncommon lesion that accounts for 0.5% of soft tissue neoplasms.[1] It occurs at any age but is most common in the black population in the 4th to 6th decades of life, with a higher female predilection.[1,2,5-7] This observation aligns with the present case: a black female in her fifth decade of life. The female-to-male ratio is 2-3:1, and the incidence in the black population is 3:1.[2,7] This lesion presented as a solitary 3 x 3cm mass on the tongue, located in the middle third of the right half of the tongue, corroborating previous studies [1,2,6,7] that the tongue is the most frequent intraoral location, followed by the soft and hard palates. The incidence of GCT on the tongue is 25%, trailing that of the skin (30-45%).[1,6] Other sites of occurrence include the breast, respiratory tract, thyroid gland, urinary tract, central nervous system, and female genitalia.[1,6]

The aetiopathogenesis of GCT is not fully established, but proposed theories include mutations in ATP6AP1 and ATP6AP2, syndrome-linked mutations, and mutations in non-neural tumor cell lineages.[2] The plausible theoretical explanation for the origin of GCT in this case (although the genetics were not investigated) may be a complex biochemical pathway within Schwann cells involving mutations in ATP6AP1 and ATP6AP2, driving sporadic tumorigenesis in the tongue musculature, as this case was

neither syndrome-associated nor a non-neural tumor derivative.[2] Besides, it reacts positively to S-100 and CD68 immunohistochemical stains.[1,2,7]

To the best of our knowledge, this case is the first GCT of the tongue reported in the Nigerian population, hence its rarity. GCT is primarily an adult lesion that reacts positively to the following IHC stains: CD68, CD57, S-100 protein, inhibin, calretinin, TFE3, SOX10, CD56, PGP9.5, and vimentin.[2,8] A similar lesion, however, the congenital granular cell tumor (CCGT), is common in neonates, which shows no immunohistochemical reactivity to S-100 protein, CD68, CD31, chromogranin A, actin, and keratin, but is positive for vimentin and neuron-specific enolase.[8] Furthermore, GCT could be confused clinically with a chondroma that occurs on the anterior two-thirds of the tongue, as both present around the 4th decade of life.[9] However, evaluating the lesions with tools like histopathology and immunohistochemistry will help distinguish one from the other.

Histopathologically, this case showed an ill-defined tumor comprising a syncytial proliferation of ovoid cells with prominent eosinophilic granular cytoplasm and regular nuclei. In some areas, the tumor entraps skeletal muscle bundles, and the overlying squamous epithelium displays pseudoepitheliomatous hyperplasia. This pseudoepitheliomatous proliferative change is seen in more than 50% of GCT cases and may be confused with squamous cell carcinoma.[2,6] The histopathological features in this case corroborate the pathognomonic features of GCT reported by Neelon et al[2] and Sposto et al.[7]

In this resource-scarce scenario, S-100 and CD68 immunohistochemical stains were used, with positive reactivity. These IHC stains confirmed the diagnosis of GCT and suggested a Schwann cell origin, consistent with prior studies.[1,5,7] GCT can also present as a malignant lesion, but the absence of nuclear pleomorphism, nuclear atypia, and mitosis helped rule out any malignancy in this case.[1,2,3,6]

Treatment is primarily surgical excision, which is usually curative and rarely recurs in benign cases.[7]

Conclusion

GCT is a rare tumour that can resemble other tongue swellings clinically, but histopathological and immunohistochemical evaluations are essential to make a definitive diagnosis.

Declaration of patient consent

The authors certify that we obtained appropriate patient consent for the publication of the patient's images and other clinical information in the journal. The patient was informed that her identity would be concealed as much as possible.

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Conflicts of Interest

Nil

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