ORIGINAL RESEARCH PAPER

Oral and Maxillofacial Pathology

ABRIKOSSOFF'S TUMOR OF THE TONGUE: REPORT OF TWO CASES AND REVIEW OF THE LITERATURE

KEY WORDS: Abrikossoff tumor, Granular Cell Tumor, Tongue

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ABSTRACT

Abrikossoff's tumor, also known as Granular Cell Tumor (GCT), is a rare benign soft tissue neoplasm of presumed Schwann cell origin that commonly affects the tongue. Clinically, it presents as a slow-growing, painless nodular swelling and may resemble reactive lesions such as traumatic or irritation fibroma. Histopathological examination, supported by immunohistochemistry, is essential for definitive diagnosis. This article reports two cases of Abrikossoff's tumor involving the tongue in middle-aged and elderly female patients presenting with asymptomatic lingual swellings. Histopathological examination in both cases revealed pseudoepitheliomatous hyperplasia of the overlying stratified squamous epithelium with underlying sheets and nests of large polygonal cells containing abundant eosinophilic granular cytoplasm and eccentrically placed vesicular nuclei. Immunohistochemical positivity for S-100 protein in one case supported the diagnosis. Complete surgical excision was performed in both cases, with no recurrence observed during follow-up. These cases emphasize the importance of considering Granular Cell Tumor in the differential diagnosis of tongue lesions because of its nonspecific clinical appearance and histopathological resemblance to malignancy.

INTRODUCTION

Abrikossoff's tumor, also known as Granular Cell Tumor (GCT), is a rare benign soft tissue neoplasm first described by Alexei Ivanovich Abrikossoff in 1926. Initially termed "granular cell myoblastoma" because of its presumed myogenic origin, it is now believed to arise from Schwann cells [1]. Clinically, GCT commonly presents as a slow-growing, painless sessile nodule covered by intact mucosa. Although it may occur in various anatomical sites including the skin, breast, gastrointestinal tract, respiratory tract, and nervous system, nearly 45%–65% of cases involve the head and neck region, particularly the tongue [2].

The tumor predominantly affects females in the fourth to sixth decades of life. Due to its nonspecific clinical presentation, the differential diagnosis includes traumatic fibroma, lipoma, neuroma, neurofibroma, schwannoma, vascular lesions, dermoid cyst, and pleomorphic adenoma. Histopathological examination is essential for definitive diagnosis. Surgical excision with adequate margins remains the treatment of choice, and recurrence is uncommon following complete removal [2].

CASE REPORTS

Case 1

A 60-year-old female presented with a painless swelling on the left lateral dorsal surface of the tongue for 3 weeks. She was a known hypertensive under medication. Intraoral examination revealed a solitary, pedunculated, firm, non-tender swelling measuring approximately 1 × 1 cm with normal overlying mucosa. No bleeding, discharge, or paresthesia was noted. A provisional diagnosis of fibroma was considered. Hematological investigations were within normal limits, and excisional biopsy was performed. Histopathological examination showed pseudoepitheliomatous hyperplasia of the overlying stratified squamous epithelium and underlying nests and islands of large polygonal cells with abundant eosinophilic granular cytoplasm and eccentrically placed pale vesicular nuclei within a moderately collagenous stroma, suggestive of Granular Cell Tumor (Figure 1). Complete surgical excision was carried out. Follow-up at 1 week, 1 month, 3 months, and 6 months showed no recurrence.

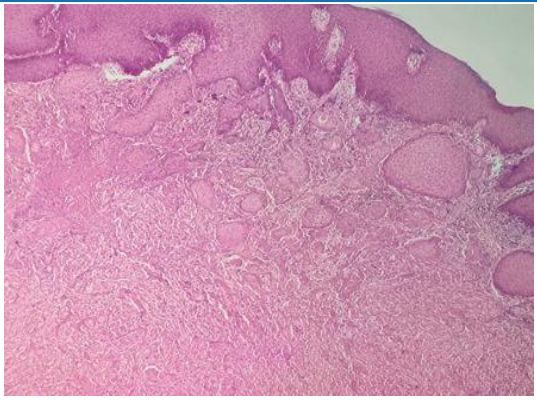

Figure 1: Photomicrograph showing pseudoepitheliomatous hyperplasia of the overlying stratified squamous epithelium with underlying sheets and nests of large polygonal granular cells exhibiting abundant eosinophilic granular cytoplasm and eccentrically placed vesicular nuclei within a collagenous stroma, suggestive of Granular Cell Tumor (H&E stain).

Case 2

A 47-year-old female presented with a painless swelling on the left side of the tongue of 4 months' duration, with a history of accidental tongue bite. Her medical history was significant for diabetes mellitus and hypertension under medication. Clinical examination revealed a solitary, firm, non-tender swelling measuring 3 × 3 cm with normal overlying mucosa and no associated bleeding, discharge, or paresthesia. A provisional diagnosis of traumatic fibroma was made. Routine hematological investigations were normal, and excisional biopsy was performed. Histopathology revealed hyperplastic parakeratotic stratified squamous epithelium overlying connective tissue composed of sheets of granular cells with abundant eosinophilic granular cytoplasm and eccentrically placed pale vesicular nuclei (Figure 2). Immunohistochemistry showed S-100 positivity, confirming the diagnosis of Granular Cell Tumor (Figure 3). Complete surgical excision was performed, and no recurrence was observed during the 3-month follow-up period.

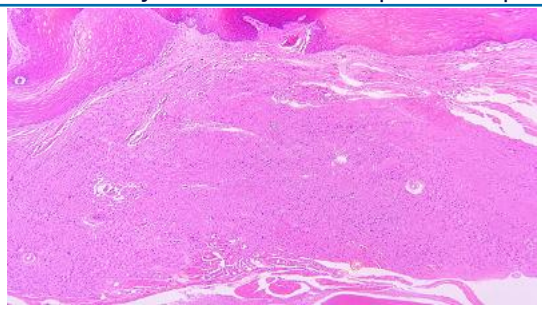

Figure 2: Photomicrograph showing hyperplastic parakeratotic stratified squamous epithelium overlying connective tissue composed of sheets of granular cells with abundant eosinophilic granular cytoplasm and eccentrically placed pale vesicular nuclei., suggestive of Granular Cell Tumor (H&E stain).

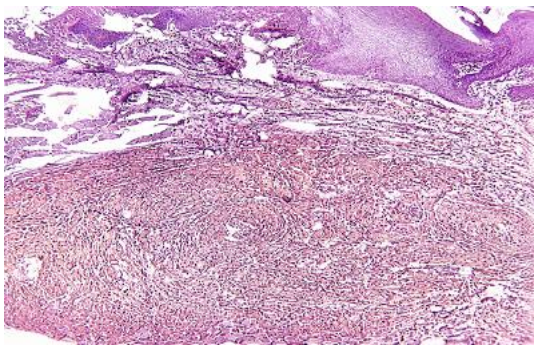

Figure 3: Immunohistochemical photomicrograph showing diffuse cytoplasmic positivity for S-100 protein in granular tumor cells, supporting the diagnosis of Granular Cell Tumor.

DISCUSSION AND LITERATURE REVIEW

Abrikossoff's tumor, also known as Granular Cell Tumor (GCT), is a rare benign neoplasm commonly affecting the head and neck region, particularly the oral cavity. It was first described by Alexei Ivanovich Abrikossoff in 1926 in a tongue lesion and was initially considered to be of myogenic origin, termed "granular cell myoblastoma" due to its resemblance to skeletal muscle fibers. However, the histogenesis of GCT remains controversial. Various origins, including striated muscle, histiocytes, fibroblasts, myoepithelium, and neural tissue, have been proposed, with Schwann cell origin now being the most widely accepted [3].

The neural origin theory is supported by the close association of GCT with peripheral nerve fibers, ultrastructural evidence of myelin figures and axon-like structures, and immunohistochemical positivity for S-100, neuron-specific enolase, and myelin proteins [4]. Although most granular cells show strong S-100 positivity, some GCTs of the lip and neck have been reported to be S-100 negative [5]. Neurogenic origin is further supported by positivity for vimentin, CD-68, PGP9.5, and NSE.

GCT commonly occurs during the fourth to sixth decades of life and shows a female predilection. Although it may arise anywhere in the body, nearly 70% occur in the oral cavity, most frequently on the tongue. About 48% occur on the dorsal surface, 15% on the lateral border, and the remainder on the ventral surface [6]. Other sites include the buccal mucosa, palate, lips, gingiva, uvula, parotid gland, skin, gastrointestinal tract, respiratory tract, nervous system, and reproductive organs. Clinically, oral GCT usually presents as a solitary, painless, firm, slow-growing pink nodular lesion measuring less than 2 cm and covered by intact mucosa. Differential diagnoses include fibroma, lipoma, neuroma, neurofibroma, and pleomorphic adenoma of minor salivary glands [7].

Histopathological examination remains the gold standard for diagnosis. Microscopically, the lesion consists of sheets of polygonal granular cells with abundant eosinophilic granular cytoplasm and vesicular nuclei. Pseudoepitheliomatous hyperplasia or acanthosis of the overlying epithelium, seen in nearly 50% of cases, may mimic squamous cell carcinoma and result in misdiagnosis [8]. CT or MRI helps determine the exact extent and localization of the lesion.

Most GCTs are benign, while malignant transformation is rare. Malignant GCT has two variants. The first shows relatively benign histology with increased mitotic activity and mild nuclear pleomorphism; hence, rapid growth, large size, ulceration, and invasion are important diagnostic clues. The second variant demonstrates progression from typical granular cells to pleomorphic granular, spindle, and giant cells with numerous mitoses. Histologic features suggestive of malignancy include necrosis, spindling, vesicular nuclei with prominent nucleoli, >2 mitoses/10 HPF, high nuclear-cytoplasmic ratio, and pleomorphism. Tumors exhibiting three or more of these features are considered malignant and carry nearly 40% mortality risk [8].

Conservative surgical excision remains the treatment of choice, and recurrence after complete removal is uncommon.

CONCLUSION

Reported cases of Abrikossoff's tumor are limited due to its rarity, with an estimated prevalence of 0.019–0.03% among all human neoplasms . Despite this, granular cell tumor should be included in the differential diagnosis of oral lesions, particularly those affecting the tongue. Although it is generally a well-recognized entity, its histogenesis and biological nature remain uncertain. Malignant transformation is rare; however, to minimize the risk of local recurrence, complete surgical excision with thorough histopathological evaluation supported by immunohistochemical analysis is recommended, along with careful long-term follow-up.

REFERENCES

1. Sargenti-Neto, S., Brazão-Silva, M. T., Nascimento Souza, K. C. do, Faria, P. R. de, Durighetto-Júnior, A. F., Loyola, A. M., et al. (2009). Multicentric granular cell tumor: Report of a patient with oral and cutaneous lesions. British Journal of Oral and Maxillofacial Surgery, 47(1), 62–64. https://doi.org/10.1016/j.bjoms.2008.06.015
2. Ottoman, B. A. (2015). Granular cell tumor of the tongue: A case report with emphasis on the diagnostic and therapeutic proceedings. 1(1).
3. Sena Costa, N. C., Bertini, F., Carvalho, Y. R., Almeida, J. D., & Rodrigues Cavalcante, A. S. (2012). Granular cell tumor presenting as a tongue nodule: Two case reports. Journal of Medical Case Reports, 6(1), 56. https://doi.org/10.1186/1752-1947-6-56
4. Vered, M., Carpenter, W. M., & Buchner, A. (2009). Granular cell tumor of the oral cavity: Updated immunohistochemical profile. Journal of Oral Pathology & Medicine, 38(2), 150–159. https://doi.org/10.1111/j.1600-0714.2008.00725.x
5. Lee, M. W., Won, C. H., Chang, S. E., Lee, M. W., Choi, J. H., Moon, K. C., & Koh, J. K. (2002). S-100-negative atypical granular cell tumor: Report of a case. International Journal of Dermatology, 41(12), 842–844. https://doi.org/10.1046/j.1365-4362.2002.01373_2.x
6. Daniels, J. S. M. (2009). Granular cell tumour of tongue: A case report. The Saudi Dental Journal, 21(2), 75–78. https://doi.org/10.1016/j.sdentj.2009.07.004
7. Goel, S., & Goel, M. (2017). Granular cell tumour of the tongue: Report of a case. Journal of Clinical and Diagnostic Research, 11(6), ZJ01–ZJ02. https://doi.org/10.7860/JCDR/2017/28946.10006
8. Barca, I., Cordaro, R., Giudice, A., & Cristofaro, M. G. (2020). Abrikossoff's tumor of the tongue: Report of three cases and review of the literature. Journal of Oral and Maxillofacial Pathology, 24(Suppl. 1), S101–S105.