

Granular cell tumor of the tongue in a 6-year-old girl- A case report

Praveen Birur Nagaraj ¹, Ravikiran Ongole ², Balaji Rao Bhujanga Rao ³

(1) Reader, Dept. of Oral Medicine & Radiology. K.L.E. Society's Institute of Dental Sciences

(2) Asst. Professor, Dept. of Oral Medicine & Radiology. Manipal College of Dental Sciences

(3) Professor and Head, Dept. of Oral Medicine & Radiology. K.L.E. Society's Institute of Dental Sciences. Mangalore. India

Correspondence:

Dr. Ravikiran Ongole

Manipal College of Dental Sciences

Light House Hill Road

Mangalore- 575001

INDIA

E-mail: oralcare@gmail.com

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ABSTRACT

Granular cell tumor is a relatively uncommon benign hamartomatous lesion occurring in almost any part of the body. The tongue and the buccal mucosa are common intra oral sites. Granular cell lesions may be found in other diverse sites such as the jaw, skin, gastro intestinal tract and respiratory tract. The histogenesis of the lesion still remains unknown. However, histochemical and ultra structural studies propose the origin of the lesion from schwann cells, striated muscle, mesenchymal cells, histiocytes and epithelial cells. The tumor generally occurs in middle or older aged adults. The lesion is typically seen as an uninfamed asymptomatic mass measuring about two cms in diameter with a yellowish surface coloration. As most of the granular cell tumors are benign, surgical excision of the lesion is the treatment of choice. We describe a case of granular cell tumor of the tongue in a 6 year old girl along with a brief review of literature on granular cell tumors.

Key words: Granular cell tumor, granular cell myoblastoma.

INTRODUCTION

Granular cell tumor (GCT) was first described by Abrikosoff in 1926 and postulated a myogenic origin and termed it as a granular cell myoblastoma (1). However the muscular origin proposed by Abrikosoff is no longer considered likely hence the term granular cell tumor is preferred to granular cell myoblastoma (2). Myoblasts (3), histiocytes (4), fibroblasts (5), undifferentiated mesenchymal cells and schwann cells (2) have been implicated in the histogenesis of GCT.

CASE REPORT

A 6-year-old girl presented with a one-year history of a painless swelling in the right lateral margin of the tongue. Two months prior to the present evaluation enameloplasty was done to smoothen the sharp edges of the teeth on the

right side. The patient reported of no regression in the size of the swelling following the grinding of the sharp edges of teeth. Clinical examination revealed a single, firm nodular mass about 3 cms in size with a yellowish white cast on the right lateral margin of the tongue. It was mildly tender on palpation. A differential diagnosis of a vascular lesion of the tongue, pleomorphic adenoma of the minor salivary glands of the tongue, dermoid cyst, lipoma, traumatic fibroma was given.

Fine needle aspiration cytology revealed groups of cells with abundant granular cytoplasm lying amidst muscle bundles. Minimal nuclear pleomorphism was noticed. Benign squamous cells to a small extent were noted. A CT scan of the neck with contrast medium revealed a soft tissue mass in the right posterolateral aspect of the tongue showing mini-

mal enhancement. Excisional biopsy was performed which revealed histopathological features (Figure 1) consistent with a granular cell tumor. The patient is presently being reviewed at six-month intervals to evaluate for recurrence and any malignant transformation.

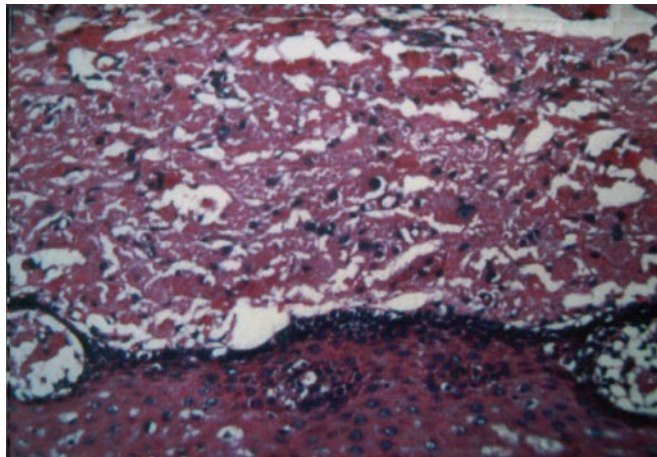


Fig. 1. Histopathological picture of the granular cell tumor showing groups of cells with abundant granular cytoplasm. Minimal nuclear pleomorphism is evident. Tumor islands are seen in close relation to the surface epithelium.

DISCUSSION

GCT is a relatively uncommon benign neoplasm that occurs in almost any part of the body such as the skin, nervous system, gastrointestinal tract, urinary bladder, female reproductive tract and bronchus (6). The head and neck region are involved in about 45 to 65% of the patients of which 70% account for intra oral lesions (7). The tongue, buccal mucosa, hard palate is commonly affected. But tumors of the lip, gingiva, uvula and parotid gland (8) have also been reported.

GCTs have been reported in patients from all age groups ranging from 11 months to 85 years (9); however the tumor most frequently occurs in the fourth to sixth decades of life and is rare in children. Females are twice as commonly affected as males (9). Benign granular cell tumors are generally seen as a solitary asymptomatic nodule less than 3 cms in size involving the subcutaneous or submucosal tissues. The mass is generally pink in color but occasionally GCTs show a yellowish surface coloration. The nodular mass is hard in consistency and generally reveals an intact overlying epithelium (10). Large lesions may sometimes show surface ulcerations, which may clinically give an impression of a malignant neoplasm. Malignant transformation of GCTs is rarely seen. An estimated 2% of the GCTs turn malignant (11). Apart from the histopathological picture, the clinical size of the tumor, pain, rapidity of growth, invasion of underlying and adjacent structures and the presence of regional and distant metastasis will aid in differentiating a benign GCT from the malignant counterpart.

Granular cell tumors exhibit round or polygonal cells with

small nuclei and abundant pale eosinophilic granular cytoplasm. The nucleus is eccentrically placed (12). The cells are usually arranged in unencapsulated sheets, but may also be found in cords and nests. The cell borders are generally indistinct giving rise to a syncytial appearance. Sometimes the tumor appears to infiltrate the adjacent connective tissue. On occasions there appears to be a transition from normal adjacent skeletal muscle fibers to granular tumor cells, this finding has led to the proposition of muscle origin for this tumor. Less frequently groups of granular cells may be seen enveloping small nerve bundles. Mitotic figures are rarely found. In lesions involving the tongue the pseudoepitheliomatous hyperplasia may be so pronounced that it has been misinterpreted as squamous cell carcinoma (10, 12).

Immunohistochemical studies of granular cell tumors suggest a neural or neuroectodermal origin of the granular cells. The neurogenic origin is supported by the immunohistochemical localization of neuron specific enolase (13) and S-100 protein markers in the tumor cells (14). A majority of the tumors follow a benign clinical course; therefore the treatment of choice is a conservative surgical excision of the lesion (6, 15). However as the GCT has a poorly defined margin it is suggested that the tumor should be excised along with portions of adjacent tissue. A low rate of recurrence of the lesion has been reported (9). Radiation and chemotherapy are not recommended because of the resistance of the tumor and potential carcinogenic effects (9, 11). A strict follow up is mandatory in all cases to rule out recurrences and to evaluate for malignant transformation.

CONCLUSION

We report an uncommon case of GCT involving the tongue in a 6-year-old girl. As granular cell tumors are unusual in the first decades of life we feel that the lesion should be included in the differential diagnosis of tumors of the tongue of young patients, among minor salivary gland tumors, vascular lesions, lipoma, benign mesenchymal neoplasm, neurofibroma, traumatic fibroma. Ultrasound imaging, CT scan and MRI may help in determining the exact location and extent of the lesion. However histological examination including antibody anti S-100 protein will help in arriving at a definitive diagnosis. We recommend that the patient be evaluated at periodic intervals to rule out malignant transformation and late recurrences.

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