Case Report:

Granular cell tumor in a young girl

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ABSTRACT:
Granular cell tumor (GCT) is a rare benign tumor of the head and neck region that commonly affects oral cavity but can also occur at other sites. The biological nature and histogenesis of the lesion is still unknown. Most widely accepted hypothesis is that GCT arises from the altered metabolism of schwann cells. We reported a case of GCT in a 16 years old female who presented with nodular swelling of buccal mucosa. Excision biopsy was done with a diagnosis of benign soft tissue tumor. On histopathology diagnosis of GCT was made. Clinical and histopathological findings including immunohistochemistry is presented along with brief discussion of literature.

Keywords: Granular cell tumor, granular cell myoblastoma, oral neoplasms

INTRODUCTION:
Granular cell tumor (GCT) is an uncommon benign neoplasm of unknown etiology. It was first described by Abrikossoff in 1926 and postulated a myogenic origin, hence termed it as a granular cell myoblastoma or Abrikossoff tumor. Subsequently various theories were proposed on the origin of GCT, including its origin from striated muscle, histiocytes, fibroblasts and a neural origin.

Granular cell tumors can affect any organ or region of the body. Most GCTs occur in the head and neck region, especially in the tongue, cheek mucosa, and palate. The only examination that can confirm the clinical diagnosis is the histological examination. We reported a case of GCT of buccal mucosa in a 16-year-old girl with immunohistochemical confirmation.

CASE PRESENTATION:
A 16 year old female presented with two-year history of painless, slowly growing swelling of left buccal mucosa of mandible. Clinical examination revealed a red single, pedunculated, well defined cystic swelling of 2cm in diameter, it was mildly tender to touch. Clinically differential diagnosis of infected cyst, vascular lesion, lipoma, traumatic fibroma were given. Ultrasonography revealed an irregular cyst approximately 1.8x1.0 cm with echogenic foci and surrounding hypoeochoic edema in muscle plane suggesting a differential diagnosis of parasitic cyst (cysticercosis). Color doppler revealed marked blood flow within tumor. Excision biopsy was performed.

Microscopic analysis revealed normal mucosal epithelium. Submucosa consisted of sheets of large round to polygonal closely packed tumor cells with abundant granular eosinophilic cytoplasm and small round central, vesicular nuclei. No nuclear atypia was observed (figure 1). The cytoplasmic granules stained positive with periodic acid-Schiff (PAS) staining and were resistant to diastase digestion. On
immunohistochemistry tumor cells were strong and uniformly positive for S-100 and NSE, and negative for desmin and vimentin (figure 2). These findings were interpreted as being sufficient to diagnose granular cell tumor.

DISCUSSION:
GCT a rare tumor is found most frequently in the head and neck region, which accounts for 45% to 65% of all sites affected by the tumor. But it can affect various other regions of the body, such as the skin, soft tissues, breast, and lungs. About 70% are located in the oral cavity, especially the tongue, oral mucosa, and hard palate. The tumor occur most often in blacks than other racial groups. GCT seems to be more prevalent among women, female to male ratio being 2:1. The tumor commonly develops between fourth to sixth decade of life and is rare in children.

The histogenesis or origin of this tumor have been matter of debate. Abrikossoff in 1926 postulated a myogenic origin and termed it as granular cell myoblastoma. In contrast to this theory others reported that approximately 50% of these tumors occurred at sites that did not contain striated muscle example gall bladder, breast and hypophysis. Some investigators favored a non-neoplastic theory and suggest inflammatory, degenerative, regenerative and congenital causes for these tumors. Azorpardi believed that GCT is a storage disease rather than a tumor. Recent IHC studies show tumor to be positive for S-100 protein thus implicating muscle or peripheral nerve sheath origin. Neurogenic origin remains the most popular explanation for these tumors and is consistent with our findings.

Benign GCT, clinically manifest as a nodular lesion that is generally asymptomatic and <3cm in size. These are smooth, sessile swelling with firm texture and involves the submucosal and subcutaneous tissues. Mucosal color varies from normal to slightly pale to yellowish in color. Overlying epithelium is usually intact, however larger lesions may reveal surface ulceration or pseudoepitheliomatous hyperplasia which may clinically mimic squamous cell carcinoma. In lesions involving tongue and laryngeal surface pseudoepitheliomatous hyperplasia can be seen.

Histologically, GCT comprises of large round to polygonal cells arranged in sheets, nests or cords. These tumor cells have abundant pale eosinophilic granular cytoplasm and small, central to eccentrically placed nuclei. Cell borders are sometimes indistinct and may give syncytial appearance. On occasion there appears to be transition from normal adjacent skeletal muscle fiber to granular tumor cells, this finding has led to proposition of muscle origin for this tumor. Malignant transformation of GCT is rarely seen. Apart from the histopathological picture, the clinical size of tumor, rapidity at growth, associated pain, invasion of underlying and adjacent structures as well as the presence of regional and distant metastasis will help in differentiating a benign GCT from malignant one.

On IHC tumor cells give strong and uniform positivity for S-100. Cells can also express NSE, CD-68, calretinin, inhibin-alpha and protein gene product 9.5 (PGP 9.5) but no reactivity for SMA. The cytoplasmic granules are PAS positive and react positively by Sudan black. Ultrastructurally, Mittal concluded that the granules were formed by infoldings of the cell membrane by a process similar to the myelin formation around axons. These infoldings were phagocytosed by lysosomes, yielding the typical granules of GCT. Based on clinical presentation, morphological features and IHC
differential diagnosis of GCT include congenital granular cell epulis, histiocytic proliferation, peripheral nerve sheath tumor, amyloidosis, lipoma, melanoma, minor salivary gland tumor and poorly differentiated carcinoma. A majority of these tumors follow a benign clinical course so treatment of choice is surgical excision. Excision should be wide as GCT has a poorly defined margin. In 15% of cases local recurrence occurs if tumor is not completely excised, whereas relapse occurs in 1-3% of cases. Radiation and chemotherapy are not recommended because of the resistance of the tumor and potential carcinogenic effects. A strict long term follow-up is mandatory in all cases to rule out recurrences and to evaluate for malignant transformation.

CONCLUSION:
Granular cell tumor is an uncommon tumor that must be carefully diagnosed and treated carefully. Even though rare in first two decades of life, the lesion should be included in differential diagnosis of tumors that arise in the oral cavity, especially when their location are the tongue or floor of mouth and patient must be evaluated at periodic intervals to rule out malignant transformation and recurrence.

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**Figure 1a:** Epithelium is unremarkable. Subepithelial tissue consist of sheets of large round to polygonal closely packed tumor cells (100x, H&E)

**1b:** Tumor cells with abundant granular eosinophilic cytoplasm and small round central, vesicular nuclei (400x, H&E).

**Figure 2:** Tumor cells showing strong and diffuse positivity for S-100 (400x).

**Figure 3:** Tumor cells showing strong and diffuse positivity for NSE (400x).
Figure 4: The cytoplasmic granules stained positive with periodic acid-Schiff (PAS) staining with diastase resistance (400x).

Figure 5: Tumor cells negative for desmin (400x)

References: