Oral Granular Cell Tumor: Report of Case Series and a Brief Review of the Literature

Introduction

The granular cell tumor (GCT) appears to be a benign neoplasm, probably of neural origin, which is usually located in the head and neck area (45% to 65% of the cases)\(^1\). Among the above, 70% of lesions are located intraorally and merely 2% of all GCTs turn out to be malignant\(^2,3,4\). A slight female prevalence is noted, especially between the third and the fourth decade of life\(^5\).

That lesion was first described by Arbikossoff in 1926, stating that the granular cells of the tumor derive from the skeletal muscles and consequently was termed myovlastoma\(^3,5\). Other theories of histogenesis claim that GCT derives from neurons, fibroblasts, mast cells and myoepithelial cells\(^6\). Furthermore, a number of authors have correlated regenerative changes and proliferation after injury with the morphogenesis of the GCT\(^6\).

According to the prevailing theory, the oral GCT derives from cells of the neural tissue, and more specifically from the Schwann cells\(^6,7,8\). In support of the last theory, the GCT is positive to the S-100 protein during the immunohistochemical examination, just as in our case\(^7,8\).

In this paper, we report a case series of 8 patients, who were examined in our clinic from 2001 to 2016.

| Table 1. Main characteristics of our GCT cases. |
|---|---|---|---|
| Age (in years) | Sex | Location | Clinical features |
| Case 1 | 7 | Female | Dorsum of the tongue | 4 mm diameter, perceptible 4 months |
| Case 2 | 55 | Male | Middle third of the right side of the tongue | 1 cm diameter, perceptible 10 years |
| Case 3 | 68 | Female | In the middle of the dorsum of the tongue | Not reported |
| Case 4 | 21 | Female | In the middle of the right edge of the tongue | 0,5 cm diameter, perceptible 6 months |
| Case 5 | 60 | Female | Top of the tongue | 0,5 cm diameter |
| Case 6 | 49 | Female | Left upper lip | 0,5 cm diameter |
| Case 7 | 60 | Female | In the middle of the dorsum of the tongue | 0,8 cm diameter, perceptible 6 months |
| Case 8 | 59 | Male | In the middle of the dorsum of the tongue | 0,6 cm diameter, perceptible 6 months, HIV+ |
The following case reports are briefly described also in Table 1. Surgical excision was the treatment of choice in all cases, presenting no recurrence. Histological examination revealed GCT.

**Case 1**
A 7-years-old girl was treated for a swelling located at the dorsum of the tongue. The swelling had a diameter of 4 mm and was being noticed 4 months before the examination.

**Case 2**
A 55-years-old male patient proceeded to our clinic for a sessile overgrowth on the middle third of the right side of his tongue (Figure 1). The lesion had a diameter of 1 cm and was being noticed 10 years before the examination.

**Case 3**
A 68-years-old female patient was diagnosed with a sessile overgrowth in the middle of the dorsum of the tongue. Further details concerning clinical characteristics of the lesion, as well as the time it appeared, were not recorded.

**Case 4**
A 21-years-old female patient was examined in our clinic and a 0.5 cm in diameter tumor-like lesion was detected in the middle of the right edge of her tongue.

**Case 5**
A 60-years-old female patient was examined for a 0.5 cm swelling on the top of her tongue. Besides histological examination, the immunohistochemical examination revealed positivity for the S-100 protein.

**Case 6**
A 49-years-old female patient was examined and a swelling of 0.5 cm in diameter was detected, located at the left upper lip.

**Case 7**
A 60-years-old female patient visited our clinic for a 0.8 cm in diameter tumor-like lesion, located in the middle of the dorsum of the tongue (Figure 2). The patient had noticed the swelling 6 months before the examination.

**Case 8**
A 59-years-old male patient was diagnosed with a GCT, found in the middle of the dorsum of the tongue. The tumor had a diameter of 0.6 cm. He had noticed the tumor 6 months before the examination (Figure 3). The patient also reported that he was HIV-positive.

**Discussion**
Eight cases of oral GCT have been described. Seven of them were located on the tongue and one at the lip. The patients’ age ranged from 7 to 60 years old. A clear female prevalence is evident since six out of eight patients were women (75%).

The oral GCT occurs at any age, although it is mainly detected in patients aged between 30 and 40 years old. The tumor might be located on any site of the oral mucosa, but the most frequent location is on the tongue, which is also confirmed by the description of our cases (87.5%).

In the case of oral GCT being reported in infants, especially in girls’ alveolar appendix, it is called congenital epulis. The unique treatment of choice is the surgical excision of the lesion. No recurrence is observed as long as the lesion has been surgically removed in toto.

The clinical examination leads to the diagnosis of the oral GCT. However, the latter should be confirmed by the histological examination (Figure 4 & 5). The epithelium covering the tumor might possibly present a pseudoepitheliomatus hyperplasia, resulting in a misdiagnosis of an oral squamous cell carcinoma. The close and substantial
cooperation between an expert pathologist and a clinician is of great importance, in order to avoid any misdiagnosis. Besides the histological examination, the immunohistochemical examination declares the positivity of the tumor for the S-100 protein, validating its neural origin (Figure 6).

Figure 4. Granular Cell Tumor. The epithelium is overlying the tumor giving a severe pseudoepitheliomatus hyperplasia appearance which could be mistaken for a squamous cell carcinoma (H-E x100)

Figure 5. Granular Cell Tumor. The cells are characterized by oval nuclei without atypia and abundant granular eosinophilic cytoplasm (H-E x400)

Figure 6. Immunohistochemistry positive for S-100 protein (magnification x400)

Conclusions

The GCT of the oral cavity is a rare benign tumor, mostly developing on the tongue. The surgical excision constitutes the treatment of choice. The histological as well the immunohistochemical examination confirms the diagnosis. As mentioned above the histological examination should be conducted carefully, especially in the presence of pseudoepitheliomatus hyperplasia, in order to avoid misdiagnosing the oral GCT as an oral squamous cell carcinoma.

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References


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