



CASE REPORT

Granular cell tumour (Abrikossoff's tumour): Case series

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KEYWORDS

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Summary This case series describes three unreported cases of an uncommon benign neoplasm named Abrikossoff's tumour or granular cell tumour (GCT). This mesenchymal neoplasm apparently arises from neural or Schwann cell origin with benign and malignant forms. All cases presented here were unique nodules on oral mucosa, coming out from the connective tissue, and occurring in women with age ranging from 30 to 42 years. The histological aspects of the GCT showed large granular cells arranged in groups and nests of connective tissue separating pseudoepitheliomatous overlying surface. This feature can possibly be misunderstood with squamous cell carcinoma. The differentiation between malignant and benign GCT is evaluated by the presence of metastases which is considered the only reliable criterion for malignancy. © 2005 Elsevier Ltd. All rights reserved.

Introduction

Granular cell tumour (GCT), Abrikossoff's tumour (AT) or Myoblastoma (My) is a rather uncommon benign neoplasm that was first described in 1926 by Abrikossoff,¹ who named it 'myoblastenmyome'. In fact, the denomination of this tumour depends on its real histogenesis, which remains unsettled, and the designation of granular cell tumour

stands as the most appropriate and more frequently used term.²

It can occur in patients of any age, although it is more common between the fourth and the sixth decades of life, being rare in children.³ GCT is two or three times more common in women than in men. Black patients are more prevalent than whites (3:1).^{2,3} Rare familial cases have been reported.⁴ Most GCTs are found in the head and neck region, and the tongue was the most common location (23–28%). GCTs have also been reported in the skin, oesophagus, larynx, stomach, bile duct, and male and female reproductive tract.^{3,5} Other sites at head and neck location have been reported including the orbit, larynx, parotid gland and

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peripheral or cranial nerves.⁶ The tumour usually presents as a single, firm, painless, small nodule in the submucous tissue, however, multiple lesions have also been reported.

The overlying epithelium frequently presents proliferative histological patterns associated or not with pseudoepitheliomatous changes. This epithelial pattern can possibly be misunderstood with squamous cell carcinoma, mainly in small fragments of tissue from incisional biopsy. The tumour cells are arranged in agglomerated strips of isolated connective tissue, which is typical feature of dense fibrous tissue. The small nuclei of GCT cells are surrounded by abundant eosinophilic cytoplasm, which resembles a granular appearance.^{2,3,5,6}

The treatment of the GCT is essentially surgical and is usually curative. The recurrence is extremely rare in these benign tumours.

The GCT can present a malignant form. However, the distinction between benign and malignant tumours is fairly difficult because there is a striking histological similarity between both and a lack of reliable criteria to provide a prediction of the tumour development.⁶

The purpose of this case series is to analyse the clinical and histological aspects in order to contribute to diagnosis understanding as well as to the treatment of benign GCTs.

Case series

Case 1

A 42-year-old woman presented to Oral Medicine Service (FOAr/UNESP) with a suspicion of squamous cell carcinoma

in the oral mucosa. She had a 26-year smoking and alcoholism history. The patient had not been aware of the lesion until her dentist performed the routine examination, when a non-tender 10 mm, firm, round, yellowish sub mucous nodule was noticed on the cheek (Fig. 1(a)). The surface was ulcerated and featured red points. Regional lymphadenopathy was not found. Surgical excision was performed and the clinical hypotheses were fibroma, neurofibroma and squamous cell carcinoma. The histological diagnosis (Fig. 2(a)–(c)) showed a granular cell tumour with margins free from illness. The follow-up was made until complete healing was achieved (1 month post-surgery) and there was no recurrence on longer follow-up (14 months post-surgery).

Case 2

A 30-year-old woman presented to Oral Medicine Service (FOAr/UNESP) with complaint of "pruride little nodule" on the tongue. She presented good systemic health and the clinical history described five months of evolution. Regional lymphadenopathy was not found. The oral examination showed a non-tender painless sub mucous nodule, 15 mm in size, (Fig. 1(b)). Surgical excision was performed and the clinical hypotheses were fibroma and granular cell tumour. The histological diagnosis showed a granular cell tumour (similar to Fig. 2(a)–(c)) with margins free from illness. The follow-up was carried out until complete healing was achieved (1 month post-surgery) and there was no recurrence on longer follow-up (13 months post-surgery).



Figure 1 (a) Nodular lesion on the right cheek with ulcerative surface (case 1); (b) nodular lesion on the right border of the tongue (case 2); (c) nodular lesion on right border of the tongue (case 3).

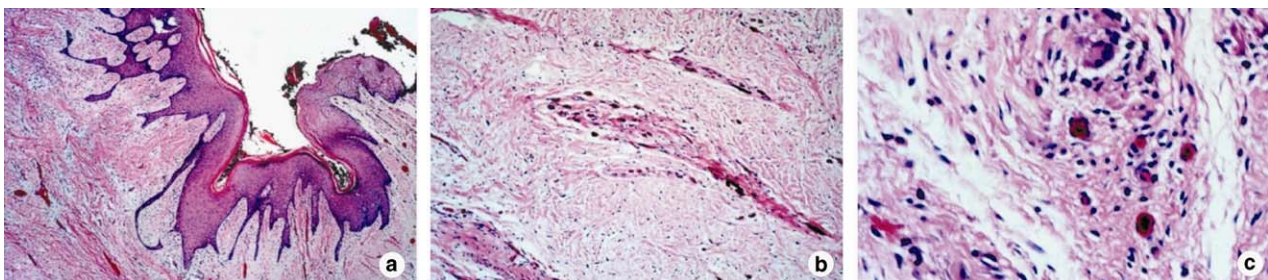


Figure 2 (a) Pseudoepitheliomatous hyperplasia of overlying and chordal granular cells separated by thin strips of connective tissue (H&E; original magnification 2×); (b) tumoral infiltration of the chorion featuring a chordal structure of granular cells (H&E; original magnification 10×); (c) agglomerations and strips of cells with abundant eosinophilic granular cytoplasm and small nuclei (H&E; original magnification 40×).

Case 3

A 42-year-old woman was referred to Oral Medicine Service (FOAr/UNESP) complaining of a mildly painful "tongue wound" growing slowly like a mass. Her past medical history showed hypertension and hypothyroidism with T4 replacement. She had been a smoker (20 cigarettes per day) for the past 15 years. She had not been aware of the lesion until the routine examination was performed by her dentist. The clinical examination showed a well-circumscribed lesion and sessile nodule, 15 mm in diameter. The mass was located just beneath the mucosa at the right lateral portion of the tongue, and had a fibrous consistency (Fig. 1(c)). The overlying non-ulcerated mucosa was intact. Surgical excision was performed and the clinical hypotheses were epulis and fibroma. The histological diagnosis was granular cell tumour (similar to Fig. 2(a)–(c)) with margins free from illness. The follow-up was carried out until complete healing was achieved (1 month post-surgery) and there was no recurrence on longer follow-up (6 months post-surgery).

Discussion

This case series shows three cases of GCT, all in women with age ranging from 30 to 42 years. The clinical appearance presented in all cases were non-tender, well circumscribed nodules on oral mucosa, varying from 10 to 15 mm. The tongue was the most affected site. All the histological analyses of the surgical specimens demonstrated margins free from illness.

GCT is usually a benign tumour and have been found throughout the body. It most frequently involves the head and neck, particularly the oral cavity (50%); the tongue and buccal mucosa are the most affected sites, respectively.^{2,5} The tumour generally occurs between the fourth and sixth decades of life and it is rare in children.³

The origin of GCT is uncertain. Abrikossoff, in 1926,¹ described a type of tumour that was originated from muscle. Later, immunohistochemical studies showed positivity for CD68 (Kp1), which allowed to classify it as a Schwannian-Origanet tumour type.⁶

In this case series, the tumours are unique, and most of the lesions are solitary nodules that arise under connective tissue. The ulcer is not usually present, however, in case 1 it was the clinical sign which led to a suspicion of squamous cell carcinoma.

Squamous cell carcinoma on the mouth arises more often at the posterior lateral portion of the tongue and may present similar clinical features. Occasionally, the CGT can mimic squamous cell carcinoma, mainly when the surface of the lesion is ulcerated due to local trauma.

The clinical aspect of the traumatic fibroma is similar to the GCT and it is the most common lesion of the oral cavity. Besides traumatic fibroma, several other possibilities must be considered when a differential diagnosis is being assessed, such as neurofibroma, schwannoma, minor salivary gland tumours, lymphomas and congenital epulis on newly-born.⁷

The histological aspects of the GCT show large granular cells arranged in groups. Strips of connective tissue separated from the surrounding areas by typically dense fibrous

proliferative tissue with pseudoepitheliomatous overlying surface, which can be confused with squamous cell carcinoma.^{2,8}

The treatment of all the cases presented here was surgical excision, and they did not show any recurrence. Surgical excision is usually an effective treatment for GCT, and recurrence is extremely rare in benign tumours.^{2,5,8}

There is little information on the results of either radiation or chemotherapy in order to determine whether these modalities are effective.⁹

It may be difficult to distinguish malignant from benign forms of GCTs through light microscopy; even sparse mitoses, mild nuclear pleomorphism, and spindling of the neoplastic cells should be reported as atypical features that raise the possibility of aggressive development. Gamboa,¹⁰ reviewing the clinical and histological characteristics of a case series of ten malignant GCTs, concluded that the malignant forms could be classified into two categories: both clinically and histologically malignant, and clinically malignant but histologically benign. Histological evidence of apparent tumoral infiltration in the tissue surrounding the tumour is a regular finding in malignant lesions.¹⁰

However, malignant forms tend to present recurrence and to metastasize within one year after the excision. It is prevalent in women (70–74%), with mean age from 40 to 48 years (ranging from 3 to 82 years).^{9,11} Malignant GCTs are uncommon, totalling less than 2% of all granular cell tumours.² In fact, 32–41% developed local recurrence, and 50–63% distant metastasis.^{9,11}

The majority of the GCTs demonstrate benign histopathologic features and also has a benign clinical course.¹² The most effective treatment for malignant GCT is the wide excision with 2–3 cm of margin. In areas where tissue preservation is essential, such as the oral cavity, Mohs' micrographic surgery may be indicated.¹³ Therefore, only clinical aspects can differentiate the malignant and benign forms of GCT, mainly through the distant metastasis. Because of this, the follow-up has been of crucial importance to GCT treatment.

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