# Abrikossoff tumor: does it origin in Swchann cells?



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# Case report

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# Abrikossoff tumor: does it origin in swchann cells? Case report

BACKGROUND: Abrikossoff tumor is an uncommon neoplasia, benign in most of the cases, that affects soft tissues, skin, and oral mucosa. Between 1% and 2% of cases are malignant and, in these cases, outcome is usually fatal. Beetween 5% and 25% of patients have multiple lesions.

CASE REPORT: A 52-year old Caucasian male was referred to the Plastic Surgery Department with a lesion in the tongue. A wide surgical excision was subsequently performed and the final pathologic diagnosis was Abrikossoff's tumor.

DISCUSSION: The pathogenesis of this tumor has long been subject of research and debate, and its origin has still to be clearly established. At the time, Abrikossoff proposed a myogenic origin, later studies supported a neural differentiation. According to the data in the literature the expression of S-100, that we found in our case, sustains the hypothesis that Abrikossoff tumor has origin in the Schwann's cells.

CONCLUSIONS: In our opinion, Abrikossoff tumor has origin in the Schwann's cells. However familial cases, congenital cases, and multiple lesions are uncommon, and it is worth stressing that this tumor has a good prognosis but every patient with Abrikossoff tumor should undergo a complete physical examination to rule out the presence of multiple associated tumors and possible visceral involvement.

KEY WORDS: Abrikossoff tumor, Granular cells, Schwann's cells.

# Introduction

Abrikossoff tumor is an uncommon neoplasia, benign in most of the cases, that affects soft tissues, skin, and oral mucosa <sup>1,2</sup>. Between 1% and 2% of cases are malignant

and, in these cases, outcome is usually fatal <sup>3</sup>. Beetween 5% and 25% of patients have multiple lesions <sup>4</sup>; moreover familial cases of multiple lesions have been reported, thus raising the possibility that some patients may have a genetic disposition towards this condition <sup>4</sup>.

Abrikossoff tumor tipically presents as a slow-growing, single, and painless nodular lesion located at the cutaneous and subcutaneous level (43%) <sup>2</sup>. It can occur in any part of the body but head and neck are more often affected respectively in 45-65% of the cases. Most of the lesions of the oral cavity show up as papule or nodule of less than three centimetres in diameter. They are asymptomatic and are generally covered by normal rapresented mucosa; but can also be verrucous <sup>5</sup>. The diagnosis in most of the cases is histopathological and treatment consists of surgical excision.

We report on a case of Abrikossoff tumor of the tongue.

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## Case report

A 52-year old Caucasian male was referred to the Plastic Surgery Department with a lesion in the tongue. The patient had no systemic complaints, on recent weight loss, and the past medical history was non contributo-



Fig. 1: Abrikossoff tumour of the dorsum of the tongue.

ry. Physical examination was unremarkable except for a  $0.5 \times 0.5$  cm solitary, hard, painless mass in the tongue dated about one year ago (Fig. 1). Blood tests and other preoperatory screening were normal. A wide surgical excision was subsequently performed. The microscopic evaluation demonstrated very characteristic granular cells, of large size, polygonal, separated by collagen, not encapsulated, with a small nucleus, abundant cytoplasm and fine eosinophilic granulations in its interior. The final pathologic diagnosis was Abrikossoff's tumor.

Immunohistochemical studies showed marked positivity for S-100 protein and vimentin.

The wound healed with an acceptable postoperative result and clinical follow-up at 8-months showed non other complication or recurrence.

# Histology findings

The Abrikossoff's tumor shows very characteristic granular cells, of large size, polygonal or fusiform, separated by collagen, not encapsulated, with a small nucleus, abundant cytoplasm and fine eosinophilic granulations in its interior. These granules are lysosomes or a component of the Golgi apparatus. Routine histopathology shows the granules to be positive for periodic acid-shift (PAS) and luxol fast blue, and resistant to diastase, indicating the presence of myelin inside the tumor (Fig. 2).

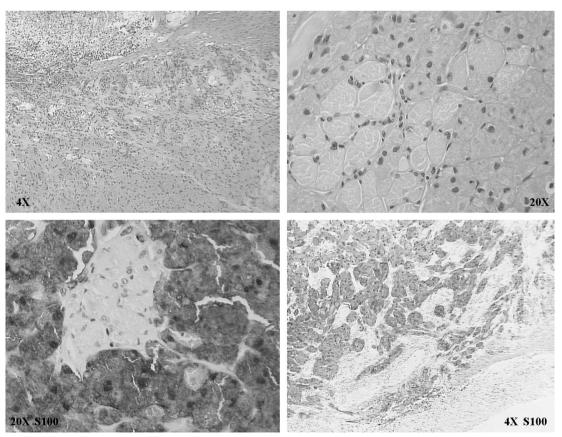


Fig. 2: Immunohistological image.

#### Discussion

This kind of tumor was first described by Abrikossoff in 1926 1. It has a typical histologic appearance that is easily recognized under optical microscope 4. However, the pathogenesis of this tumor has long been subject of research and debate, and its origin has still to be clearly established. At the time, Abrikossoff proposed a myogenic origin, deeming that the tumor was the result of cell degeneration of the striated muscle, and thus classifying it as a myoma <sup>6,7</sup>. Later studies supported a neural differentiation, the hypothesis first put forward by Feyrter in 1935 <sup>1</sup>. Through immunoenzymatic studies, other authors agree with the neurogenic origin 8. In 1962, Fisher and Wechsler used an electronic microscope to confirm that the condition could represent differentiation from Schwann's cells. This is the hypothesis currently accepted for Abrikossoff tumor 1. According to the data in the literature the expression of S-100, that we found in our case, sustains the hypothesis that Abrikossoff tumor has origin in the Schwann's cells. Vimentin suggests mesenchymal origin, however this is also rapresented in other cells, as hystiocyes, condrocytes endothelial cells, being thus unspecific.

#### Conclusions

In our opinion, Abrikossoff tumor has origin in the Schwann's cells. However familial cases, congenital cases, and multiple lesions are uncommon, and it is worth stressing that this tumor has a good prognosis but every patient with Abrikossoff tumor should undergo a complete physical examination to rule out the presence of multiple associated tumors and possible visceral involvement.

### Riassunto

BACKGROUND: Il tumore di Abrikossoff è una neoplasia rara, benigna nella maggior parte dei casi, che colpisce i tessuti molli, la cute e la mucosa orale. Nel 1-2% dei casi il tumore è maligno, e ciò è associato ad una prognosi infausta. Il 5-25% dei pazienti sviluppa lesioni multiple.

CASO CLINICO: Uomo caucasico di 52 anni si presentava alla nostra attenzione per la presenza di una lesione

sulla lingua. Veniva eseguita un'escissione chirurgica della lesione, la quale, all'esame istologico, risultò essere tumore di Abrikossoff.

DISCUSSIONE: L'eziopatogenesi del tumore di Abrikossoff è da sempre molto dibattuta. Al tempo della sua scoperta Abrikossoff propose un'origine miogenica, studi successivi suggerirono un'origine neuronale. In accordo con i dati pubblicati in letteratura, l'espressione di S-100, che abbiamo ritrovato nel nostro caso, supporta l'ipotesi che il tumore di Abrikossoff origini dalle cellule di Schwann.

CONCLUSIONI: Secondo i nostri studi, il tumore di Abrikossoff origina dalle cellule di Schwann. Inoltre forme multiple, forme familiari e congenite sono eventi rari, a conferma della buona prognosi di questo tumore. Ad ogni modo tutti i pazienti affetti da tumore di Abrikossoff dovrebbero essere sottoposti ad esame fisico completo per identificare eventuali localizzazioni multiple o possibili coinvolgimenti viscerali.

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