Oral Abrikossoff Tumor: A Report of Uncommon Presentation

Ashwini1, V. Naveen Shankar* and V. Praveena2

1Kothiwal Dental College Research Centre and Hospital, Department of Oral Medicine and Radiology, Moradabad, Uttar Pradesh, India
2Cure and Care ENT Centre, Chikkaballapura, Karnataka, India

Abstract

Abrikossoff tumor or Granular cell tumor usually present as unencapsulated, small, firm white mass, usually less than 2 cm with infiltrative borders. The histogenesis seems to be neural and most likely Schwann cell in origin. The lesion will present as an asymptomatic mass usually in the submucosa. Usually it behaves as a benign lesion; however, about 2% of the cases can present a malignant course. Here we report a case of large diffused painful swelling in the palate in a 9 year old boy. This was diagnosed as benign granular cell tumor on histopathologically, but had malignant clinical characteristics.

Keywords: Abrikossoff tumor; Granular cell tumor; Schwann cell; Granular cells

Introduction

Abrikossoff tumor, otherwise known as myoblastoma or Granular cell tumor (GCT) [1]. GCT is an uncommon benign lesion affecting the mucous membrane of the upper aerodigestive tract. About one third of all GCTs occur in the head and neck region. The most common site is the anterior part of the tongue [2]. Abrikossoff’s tumor is of uncertain origin that has been variably considered a true neoplasm, a degenerative metabolic process or a trauma-induced proliferation. The hypothesis of neural origin has been more widely accepted. However, reports suggesting a possible muscular, histiocytic, fibroblastic or pericytic origin can be found in the literature [3].

Oral Abrikossoff tumor show a predilection for females (twice as common than males), the age range varies from 4 months to 89 years, with a mean appearance in the fourth decade however they are rare in the first decade. Here is a aggressive appearing GCT clinically which turned out as benign GCT seen in 9 year old boy.

Case Report

A 9 year old boy was referred to oral diagnostician for evaluation of painful swelling in the palate since 15 days. The swelling was insidious in onset and gradually increasing in size, associated with continuous throbbing pain. He reports of no previous history of neither trauma nor pus discharge from the swelling.

On examination, a solitary swelling in the mid palate was noted measuring 3.5 X 1.5 cms extending from incisive papillae to the junction of hard and soft palate antero-posterior direction and extending 0.75 cms on either side of midline. The mucosa covering the swelling appeared normal. The lesion was slightly tender, soft in consistency and not palpable (Figure 1).

Radiographic examination revealed no changes in the odontogenic structures or in the palatal bone. Abscess, Soft tissue tumor, and salivary gland pathology was thought as differential diagnosis. Surgically the lesion was excised. Histopathologically the connective tissue stroma revealed presence of oval granular cells with abundant fine granular eosinophilic cytoplasm. These cells are separated by fibrous septa. Based on these findings the lesion was diagnosed as benign granular cell tumor (Figure 2). The patient was followed for 6 months without any relapse and he is on further follow-up.

Discussion

The GCT was first described in 1926 by a Russian pathologist Aleksei Ivanovich Abrikossoff (1875–1955), in a patient with a lesion of the tongue. He classified it as having a myogenic origin [1]. In 1931, due to the finding of analogies between myoblasts and granular cells of the tumour, Abrikosov called it as myoblastoma [4]. Williams et al. [5] suggested a neuroendocrine origin of the tumour, based on immunohistochemical results. This theory was sustained by Holland et al. who demonstrated positivity for S-100 in neurons and Schwann cells, but not in muscular fibres [5].

The neoformation can affect all parts of the body. The head and neck areas are affected in 45% to 65% of cases and of these, 70% are located interorally (tongue, gingival and oral mucosa) of which 67–81% of the lesions occurring in the tongue [1,7]. In the present case the...
lesion was involving the hard palate which is the uncommon site for Abrikosoff tumor. Incidence of GCT in hard palate is unknown [2].

Generally GCT’s are due to the response of the tissue to many factors, such as: neoplasia, degenerative or reactive processes like anaemia, metabolic disorders and lysosomal effects [3]. Abrikosoff tumor in the oral cavity was attributed to chronic trauma exercised by the teeth on the oral mucosa. The tumor presents as papule or nodule, varying in dimensions from 2 to 5 cm, causing no pain. The mucosa that covers it is of normal appearance, although it can vary in color from white to grey or blue. The tumor is not encapsulated but is well removable by the surrounding tissue [1,2,4]. In the present case the lesion did present as swelling in the palate with normal mucosa covering the lesion. But the lesion was measuring about more than 3cms and it was associated with pain these features were in contrast to the benign nature of the lesion.

Malignant form of GCT can occur which ranges from 1% to 3%. The malignancy is largely suggested by its rapid growth, the broad dimensions (>4 cm), and the presence of necrotic and hemorrhagic areas [8]. In 1998, Fanburg-Smith et al. [9] suggested six histological criteria for malignancy and prognosis of Abrikosoff tumor (Table 1). Neoplasia presenting three or more of these criteria are classified as histologically malignant, those with one or two criteria are classified as atypical; those presenting only focal nuclear pleomorphism, without any additional criterion, are classified as benign [9]. In the present case, granular cells were seen deep in the stroma separated by fibrous septa. There were no malignant histological features in our case on the contrary it had malignant clinical features.

The benign form of GCT’s are most common, shows 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 believed to be phagolysosomes, suggesting a degenerative process associated to these granular cells [1,2].

Surgical excision is the treatment of choice with local excision of the overhanging mucosa and the underlying periosteum. However, a small percentage of recurrence is described. The recurrence rate after adequate local excision is 8%. Regional lymph node dissection is recommended when tumors have rapid growth or when they exceed 4 cm in diameter. Radiation therapy and chemotherapy have been used in malignant Abrikosoff tumor. Their effectiveness is not established [8].

**Conclusion**

In the present case clinical feature were of malignant nature, whereas histopathologically the lesion had benign features. For the proper management of GCT both clinical and histopathological features must be correlated. However, regional metastasis should be ruled out with proper follow up.

**References**


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**Table 1: Malignant features of Abrikosoff tumor.**

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<thead>
<tr>
<th>Location</th>
<th>Features</th>
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<tbody>
<tr>
<td>Clinical</td>
<td>Rapid growth.</td>
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<td></td>
<td>Pain.</td>
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<td></td>
<td>Large lesions (&gt;4 cm).</td>
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<tr>
<td></td>
<td>Presence of necrotic and hemorrhagic areas</td>
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<td>Histological</td>
<td>Necrosis.</td>
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<td></td>
<td>Cells in fusiform strings.</td>
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<td></td>
<td>Nuclear pleomorphism.</td>
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<td>Large nucleus with vesicular core.</td>
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<td></td>
<td>Increased mitotic activity.</td>
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<td>Increased nucleus radius in relation to cytoplasm.</td>
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**Figure 2:** (2 X view) Oval granular cells with abundant fine granular eosinophilic cytoplasm noted in the connective tissue.