REVIEW ARTICLE

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Abrikossoff tumor of the tongue in a teenager – case report

Guz Abriksowa języka u nastolatki – opis przypadku

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ABSTRACT: Granular cell tumor is benign neoplasm rarely diagnosed among young children and adolescents. The tumor developed commonly within mucous membrane of upper airways, but precise etiology is not known. Treatment is based on surgical resection of tumor and intense follow up due to risk of recurrence and malignant transformation.

KEYWORDS: Granular cell tumor, Abrikssoff tumor, benign neoplasms, tongue

STRESZCZENIE: Guz ziarnistokomórkowy to nowotwór o charakterze niezłośliwym, występujący niezwykle rzadko w grupie dzieci i nastolatków. Do jego rozwoju dochodzi najczęściej w obrębie błony śluzowej górnych dróg oddechowych. Dokładna etiologia tego nowotworu nie jest do końca poznana. Leczenie opiera się na chirurgicznej resekcji zmiany i dalszej obserwacji chorego. Ze względu na ryzyko wznowy oraz transformacji do nowotworu złośliwego, pacjent powinien mieć częste kontrole.

SŁOWA KLUCZOWE: Guz ziarnistokomórkowy, Guz Abriksowa, nowotwory niezłośliwe, język

INTRODUCTION

Granular cell tumor is also known as Abrikossoff tumor in honor of the Russian pathologist Alexei Abrikossoff, who was the first to describe this type of lesion in 1926. This is a benign neoplasm of unknown origin, probably arising from Schwann cells. Its most common location is the head and neck, particularly the upper respiratory airway mucosa [1]. Usually, the oral cavity is affected, tongue in particular. The second most common location is laryngeal mucous membrane [2, 3]. The tumor develops in the fourth and sixth decade of life. The reports in the literature on granular cell tumors in children and adolescents are scarce [4]. Some tumors can be malignant, spreading to local lymph nodes and to distant locations. Microscopically, they show malignant features.

CASE REPORT

A 14-year-old female patient was referred to the Department of Otolaryngology at the Medical University of Warsaw for surgical treatment of a lump on her tongue. The lesion was first noticed by the patient and her parents about 12 months before admission. The patient said that over that time the tumor did not grow. She denied any pain, loss of taste, limited tongue mobility or previous trauma. On ENT examination, no other pathologies were noted except for an oval mass located at the tip of the tongue on the left side, 20 x 10 mm in size, non-tender and fixed to the tongue. On palpation, no cervical lymphadenopathy was found. The lab test results were normal, and after consultation with the anesthesiologist, she was qualified for surgical resection under general endotracheal anesthesia.

During the procedure, we performed a marginal incision of the lingual mucosa on the left side, then the mass was separated from the surrounding tissues. The tumor presented grossly as an oval cream-colored and well-bordered mass 20 mm in length. Due to difficulties dissecting the mucous membrane of the dorsum of the tongue, we decided to remove both structures together.

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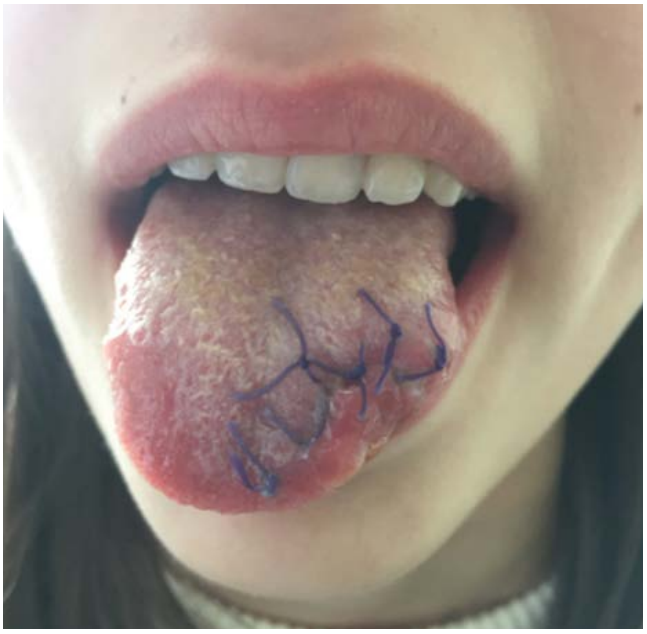

Fig. 1. The patient on the 7th postoperative day shows normal wound healing following tumor resection and local reconstruction.

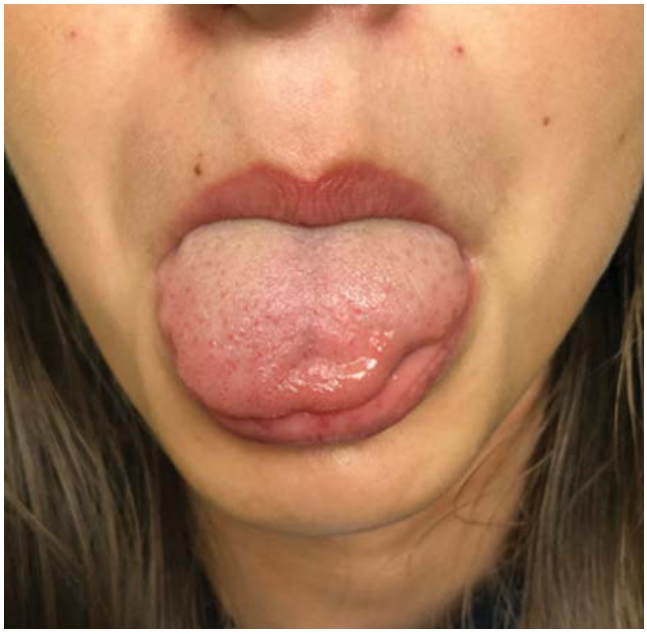

Fig. 2. The patient 6 months after surgery.

For a satisfactory esthetic and functional effect, the wound was closed after mobilization of the surrounding mucous membrane to fill in the defect (Photo 1). Postoperative period was uncomplicated. The patient negated limited tongue mobility, difficulty eating or loss of taste. On gross examination, the tumor was described as a greyish-yellow solid mass 15 x 15 x 8 mm in size, partially covered with lingual mucosa. On microscopic examination using hematoxylin and eosin (H&E) staining, polygonal cells rich in plasma were observed with centrally located nuclei and strongly eosinophilic pustulo-ovoid granules (so-called Milian bodies), indicating granular cell tumor. To confirm the initial diagnosis, additional immunohistochemical staining was performed, which was positive for S100 and CD68 proteins and PAS reaction, and negative for Caldesmon. It allowed to make the final diagnosis and exclude malignancy. Based on the pathology report, complete resection and good local postoperative condition, we decided for and planned further observation during frequent follow-up visits. Over the first year of observation, no clinical features of local relapse were noted (Photo 2).

DISCUSSION

Granular cell tumor (Abrikossoff tumor, granular cell myoblastoma) is a benign neoplasm affecting predominantly females in their 30s to 50s. It is rarely seen in children and adolescents. Until 1998, only 20 cases of Abrikossoff tumor were reported in patients younger than 17 years [4]. The etiology is not fully understood, but it is assumed that it can be associated with congenital anomalies, metabolic disorders, trauma or cancerous transformation of Schwann cells. It is estimated that multifocal synchronous primary tumors are present in 2-10% of cases [5]. The mass is located within the head and neck in 50% of cases, usually involving the tongue and upper respiratory airway mucosa. On examination, a single non-tender mass less than 3 cm in diameter can be observed. The lesion can be covered with normal mucous membrane, or it can appear pale yellow when located superficially [6].

The differential diagnosis should include non-neoplastic lesions (granulomas, amyloid deposits in amyloidosis), other benign tumors (schwannoma, paraganglioma, angioma) or malignant tumors (squamous cell carcinoma, chondroma, chondrosarcoma). For more in-depth evaluation of disease advancement, the diagnostic workup should include imaging studies. On computed tomography scans, the granular cell tumor is a solid lesion with regular contrast enhancement. On magnetic resonance imaging, the mass is hypointensive on T1-weighted scans and shows uniform contrast enhancement as well [7].

Microscopic study with H&E stain reveals round or fusiform cells containing strongly eosinophilic cytoplasm. Numerous cytoplasmic granules positive for PAS reaction are typical. To confirm the diagnosis, additional immunohistochemical staining can be performed for S-100 and CD68 proteins (positive results) or keratin, desmin, myoglobin and actin (negative results) [4, 8, 9].

Usually, small lesions are removed completely. In uncertain cases or extensive spread, fine-needle biopsy or tissue sample should be considered for cytology assessment. Due to common location on the tongue, biopsy requires preparation of the patient, physician performing the procedure and pathologist experienced in head and neck biopsy evaluation. In order to provide reliable results, it is advisable to clean and disinfect the tongue surface with hydrogen peroxide or other colorless disinfectant. The surrounding tissues should be injected with a local anesthetic, then small-dose lidocaine (1 cm3) may be considered. The biopsy should be performed using a fine needle 0.5 mm in diameter (25G), sampling material from 3 to 4 distinct sites. The material obtained in such a manner should be sufficient for a couple of smears used for various stains and further microscopic analysis. Some sample material can be placed in a fixative solution and used later for paraffin blocks, stains and microscopic study. Such a cytologic material can be used for immunohistochemical staining for more in-depth evaluation. However, paraffin block preparation is associated with a much longer preparation time before microscopic evaluation and hence more time is needed before the study report is released [10].

Multiple granular cell tumors can develop in patients with congenital LEOPARD syndrome, which is inherited in an autosomal dominant pattern. Mutations of PTPN11 gene encoding tyrosine phosphatase, leading to many abnormalities such as lentigines on the skin, bundle branch block, cardiovascular malformations (usually pulmonary artery stenosis), facial dysmorphic features (hypertelorism, prognathism, triangular face, ptosis, epicanthic fold), sensorineural hearing loss often detected in childhood, failure to thrive. In 1976, Veron et al. proposed the following diagnostic criteria: presence of lentigo and two dysmorphic features or confirmed LEOPARD syndrome in close relatives plus three dysmorphic features. Nowadays, it is believed that clinical suspicion should be confirmed by genetic study showing mutation [11, 12].

Prognosis in patients with granular cell tumor depends on its malignancy. For benign lesions, the prognosis is very good; however, mean survival in patients with malignant lesions is 44 months (95% CI 11-76 months). Malignant lesions account for about 2% and can be divided into two categories based on microscopic image and clinical course. In the first group, the microscopic image resembles that of a benign lesion, but metastases can be found in lymph nodes and distant locations including lungs, liver and bones. The second group consists of lesions with malignant microscopic image (necrotic areas, vacuolated nuclei and nucleoli, high nucleus-to-cytoplasm ratio). In the study by Aksoy et al., the authors emphasize that it is difficult to precisely determine the character of a tumor and it is necessary to observe the patient for at least two years. The authors point out to an increased expression of p53 and Ki-67 proteins in malignant lesions, which can be useful for differentiation [3, 13].

CONCLUSIONS

Granular cell tumor is a rare, usually benign neoplasm of unknown origin. It usually involves the mucous membrane of the head and neck. It is believed to arise from Schwann cells. Surgical resection is the mainstay of treatment accompanied by further long-term follow-up. The monitoring is necessary because of possible relapse after non-radical resection or malignant transformation. This case shows typical presentation of the tumor in oral mucosa but at an atypical age, which is rarely encountered.

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