Rhabdomyoma in the base of the tongue: A case report

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ABSTRACT

Introduction: Rhabdomyomas are benign and rare mesenchymal tumors. They are classified into cardiac and extracardiac. However, the majority of adult subtype, which are extracardiac, tend to occur in the head and neck region.

Present of case: A 57-year-old man with dysphagia, dysphonia and stertor one year ago. Head and neck endoscopy was the first step to detect a smooth mass at the base of the tongue, after that CT-scan showed great lobuled mass measured $7 \times 5 \times 6$ cm.

However, complete surgical excision was done and the histopathological examination play a central role to reveal an adult-type rhabdomyoma.

Discussion: the tongue base is absolutely one of the rarest sites of oral adult-type rhabdomyoma (ARM), because since 1948 to 2021 there were only four English-language articles which described ARM in the base of the tongue, anyway this type mimics malignant tumors on CT-scan because of its unclear borders and The golden diagnosis depends on histopathological examination and immunohistochemistry staining when the desmin markers are positive after that the tumor was completely excisioned as the best treatment.

Conclusion: Base of the tongue is a very rare location for adult-rhabdomyoma which must be considered in the differential diagnosis of head and neck lesions.

1. Introduction

Rhabdomyomas (RM) are benign and rare mesenchymal tumors made up of striated muscle cells. They are classified into intracardiac and extracardiac lesions. Intracardiac rhabdomyomas are considered hamartomatous lesions which are seen in infants and children and associated with phacomatoses, especially tuberous sclerosis. Extracardiac rhabdomyomas are clinically and morphologically subdivided into three types: genital, fetal and adult type. Adult-type rhabdomyoma (ARM) is the most common of all extracardiac subtypes and represents fewer than 2% of all muscular tumor. ARMs occur mostly in elderly men and have a penchant for the head and neck region like oral cavity, larynx, pharynx and soft tissue [1,2].

This paper represents a rare case of multi-lobulated ARM in the base of the tongue and the anterior part of oral pharynx.

However, this work is also reported in line with SCARE criteria which helped to improve the transparency and quality of this case report [3].

2. Case presentation

A 57-year-old male referred to the Head and Neck Surgery clinic with a complaint of one-year of dysphagia, dysphonia and stertor, with a history of two incomplete laryngoscopic operations on the inlet of the larynx. The physical examination was normal and neck lymph nodes were not palpable. Head and neck endoscopy showed a great smooth mass covered by normal mucosa at the base of tongue, the mass Pushed the larynx without affecting vocal cords movement. Computed tomography with contrast was performer and it revealed a great multi-lobulated mass measured $7 \times 5 \times 6$ cm at the anterior part of the oral pharynx and centralized in the base of the tongue, it pushed the adjacent muscles and reached the submandibular gland, epiglottis and hyoid.

Abbreviations: RM, rhabdomyoma; ARM, adult-type rhabdomyoma; CT, computed tomography.

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bone without damaging the bone or invading great vessels (Fig. 1).

The huge size of the mass imposed us to do tracheostomy instead of intubation. Subsequently, surgical excision was performed by external approach 4 cm under the left submandibular and forward to the hyoid bone, the left submandibular gland was removed and the tumor was dissected from the mucosa of oropharynx, after that we removed all the tumor and a part of the adhesive oropharyngeal mucosa to avoid recurrence, after that the mucosa of the pharynx, base of the tongue and ephyroid muscles were sutured (Fig. 2). Histopathological analysis showed a tumor measured up to 7 cm and involved submucosal skeletal muscle tissue, it composed of well demarcated lobules and nodules of sheets of large cells with abundant eosinophilic cytoplasm, focally clearing and small nuclei (Fig. 3). Immunohistochemically, desmin markers showed 100% positive staining for the tumor cells which confirmed the diagnosis of adult-type RM (Fig. 3). There were no signs of recurrence after 24 months of clinical and imaging follow-up.

Fig. 1. Computed tomography (CT) of the neck showing a large mass at the anterior part of the oral pharynx and deeply located in the base of tongue.
3. Discussion

Zenker [4] in 1864 introduced for the first time the term “rhabdomyoma”, which was used to describe a benign tumor consisting of striated muscle cells with various degrees of maturation and differentiation.

ARM usually occurs as a solitary lesion (70%), multinodular (26%) and rarely multicentric (4%). It is mostly reported in men over 50 years of age, head and neck are the most common site for ARMs followed by the extremities, esophagus, stomach, mediastinum and orbit. ARMs are considered rare in the oral cavity and there are about 80 cases which have been reported in the English-language literature. However, the tongue base is absolutely one of the rarest sites of oral ARM, whereas since 1948 to 2021 there were only four English-language articles described ARM in the base of tongue [5]. The first clinical manifestations are non-specific for ARMs and depend on the size and site of the lesion, such as: painless, globus sensation dysphagia and hoarseness [6]. Our patient came with a history of dysphagia, dysphonia and stertor one year ago.

Imaging usually shows an absence of invasion of the surrounding soft tissues and the submucosal location, which refer to benign lesion, but on CT-Scan borders that blend into adjacent isodense muscles are unclear in the adult-type RM, which mimic malignant tumors [6,7]. The final and gold diagnosis for adult rhabdomyoma depends on histopathological examination and immunohistochemistry staining [6]. In our case the pathology result showed adult type RM composed of sheets of large round cells with abundant eosinophilic cytoplasm.

The treatment of choice for RM is complete surgical excision, which can be done either by endoscopic or external approach. However, the tumor was huge (about 7 cm) blocking the entrance of larynx so that the endotracheal intubation wasn’t possible. Unfortunately, the rate of recurrence of AR after excision has been reported to occur about 16% to 20% especially in case of incomplete excision [5,6]. There is no malignant transformation has been described.

4. Conclusion

Adult-type RM in the base of the tongue is a really rare benign tumor
and one of the differential diagnosis of head and neck lesions. Although the radiological imaging couldn’t differentiate the rhabdomyoma clearly, so we get to the final diagnosis by histopathological study after the complete surgical excision.

**Availability of data and materials**

All data are available from the corresponding author on reasonable request.

**Provenance and peer review**

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**Ethical approval**

This study has been approved by the ethical committee of Damascus University.

**Consent**

Written patient consent was obtained before reporting this case and before using any of his medical information or surgical pictures.

**Author contribution**

Bairak Salameh: Reviewed the literature, wrote the article’s abstract, introduction, part of discussion, and designed the figures and provided all the captions of the figures.

Amjad Ghareeb: wrote the introduction, and part of the discussion provided the language - editing services.

Hadeel Badran: Reviewed the literature, wrote the case presentation.

Hayan Salameh: Reviewed the literature, wrote the case presentation with Hadeel and edited the figures.

Wahib Hajali: wrote the conclusion and reviewed the literature.

Areej Alassaf: Led the surgery and supervised the scientific and academic aspects of the manuscript preparation and submission. All the authors read and approved the content of this manuscript.

**Registration of research studies**

Not applicable.

**Guarantor**

Mr. Bairak Salameh.

**Declaration of competing interest**

All the authors declared that they have no conflicts of interest.

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