Rhabdomyoma of the Parapharyngeal Space – A Case Report

Rhabdomyoma przestrzeni przygardłowej – opis przypadku

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ABSTRACT: Rhabdomyomas (RM) are rare benign mesenchymal tumors. They are a much more uncommon entity than their malignant counterparts, rhabdomyosarcomas. Rhabdomyomas fall into two general categories: cardiac tumors and extracardiac tumors. Extracardiac myomas are among the rarest tumors in humans and can be classified as fetal and adult, depending on the degree of differentiation of individual tumors by light microscopy. Adult extracardiac myoma is most commonly characterized by the occurrence in the head and neck, and mainly in the laryngeal and pharyngeal area.

Case report: This article presents a case of rhabdomyoma of the parapharyngeal space with a review of world literature.

KEYWORDS: benign mesenchymal tumor, dysphagia, parapharyngeal space, rhabdomyoma

STRESZCZENIE: Mięśniaki prążkowanokomórkowe (łac., ang. rhabdomyoma; RM) to łagodne guzy mezenchymalne, zwłaszcza w porównaniu z ich złośliwą formą – mięśniakomięsaki prążkowanokomórkowymi. Są dużo rzadszą jednostką chorobową niż ich złośliwe odpowiedniki – mięśniakomięsaki prążkowanokomórkowe. Rhabdomyoma dzieli się na dwie kategorie: guzy serca i guzy pozasercowe. Mięśniaki pozasercowe należą do najrzadszych nowotworów u ludzi i można je zaklasyfikować jako formę płodową lub dorosłą, w zależności od klinicznych i morfologicznych różnic oraz stopnia zróżnicowania. RM pozasercowy typu dorosłych występuje najczęściej w rejonie głowy i szyi, głównie w obszarze krtani i gardła.

Opis przypadku: W artykule przedstawiony został przypadek mięśniaka prążkowanokomórkowego przestrzeni przygardłowej wraz z przeglądem literatury światowej.

SŁOWA KLUCZOWE: dysfagia, łagodny guz mezenchymalny, mięśniak prążkowanokomórkowy, przestrzeń przygardłowa

ABBREVIATIONS

ARM – extracardiac rhabdomyoma – adult form
ATR – extracardiac rhabdomyoma – adult type
CR – cardiac rhabdomyoma
ER – extracardiac rhabdomyoma
FRM – extracardiac rhabdomyoma – fetal type
GRM – extracardiac rhabdomyoma – genital type
MR – magnetic resonance imaging
RM – rhabdomyosarcoma

INTRODUCTION

Rhabdomyomas are among the rarest benign tumors in the human population. Histologically they derive from the striated muscle and in view of their location they are classified as cardiac rhabdomyoma (CR) and extracardiac rhabdomyoma (ER). The first type is often linked with genetic disorders, supposedly of a developmental nature of hamartoma type, and appear almost exclusively in the hearts of infants. Based on clinical and morphological differences and the degree of differentiation, extracardiac rhabdomyoma is divided into different groups: a rare fetal type (FRM), a more common adult type (ARM), which mainly affects the head and neck, as well as the genital type (GRM), which appears in the vulva and vagina of women [1]. Adult ERs are rare. They are morphologically characteristic, benign mesenchymal tumors with high skeletal muscle cell diversity. In 90% of cases, they affect the head and neck, mainly in the mucous membranes of the oral cavity, the pharynx, nasopharynx and larynx. The mean age of patients is 60 years, with a dominance of men [2]. We present a case of rhabdomyoma of the pharyngeal space and discuss misleading clinical findings that have led to inaccuracy in the preoperative diagnosis.
A 73-year-old patient was referred to the Clinic of Otorhinolaryngology of Head and Neck Surgery of the Medical University of Warsaw from another center for surgical treatment. The patient was in good general condition upon admission. Despite dysphagia persisting for 7 years, he did not report any other conditions. A year prior to admission, imaging diagnostics (head MRI) performed due to neurologic recommendation accidentally revealed a tumor of the right palatine tonsil, involving the right pharyngeal space. To deepen diagnostics, specimens of the palatine tonsil and nasopharynx on the right side were collected three times. Each result of histopathological examination excluded a malignant tumor.

Physical examination revealed a prominent right palatine arch with an enlarged right palatine tonsil, without coating or ulcerations. During palpation, the tonsil was soft in consistency. It was recommended to repeat the MR of the head and neck, which revealed a 42 x 25 x 50 mm lesion, involving the right palatine tonsil and the right lateral wall of the oropharynx. Following radiological consultation, it was decided to perform right-sided tonsillectomy due to suspected presence of a tumor in the palatine tonsil. The surgery was uneventful; the tonsil was excised without difficulty, and it did not appear cancerous. The postoperative course was uneventful as well – the patient was discharged home on the 5th day and recommended to report for follow-up and for histopathological examination in 3 weeks. Histopathological examination of the tissue material did not reveal malignant infiltration of surrounding tissues, focal lesions or dysplasia.

Follow-up examination after 3 weeks from surgery revealed uvula shifted to the left and the presence of protrusion of the lateral pharyngeal wall on the right. MR of the head and neck was repeated. The result of the examination was radiologically consulted. The current images were compared with previous pictures, and it was found that the tumor mass filled the right pharyngeal space. This was not evident on MR prior to tonsillectomy. The previous image was interpreted as a displacement of the lateral wall of the pharynx by the tonsil mass. Under these conditions, a decision was made to excise the tumor of the right pharyngeal space. Both the surgery and the postoperative period progressed uneventfully and the patient was discharged home in good general condition. Histopathological examination revealed a mesenchymal tumor of adult-type rhabdomyoma.

DISCUSSION

Extracardiac rhabdomyomas are very rare benign neoplasms and constitute less than 2% of striated muscle tumors [3]. In 1987 Pendell was the first to describe this disease entity, presenting the case of a fetal rhabdomyoma, and by the beginning of the 21st century, fewer than 100 cases of this tumor were described in world literature [4, 5]. Extracardiac rhabdomyomas most often involve the head and neck, and develop from the muscles of the third and fourth branchial arches. Helliwell described the most common locations of ATR for the head and neck, with a dominance of extracardiac rhabdomyomas in the larynx. Furthermore, it has been reported to occur in the orbit, lip, floor of the mouth, tongue, submandibular glands, soft palate, pharyngeal space and esophagus [5, 6, 7].

ARM is observed in adults in the 6th–7th decade of life and is more frequent in men than in women, with a 3–5:1 ratio [2, 5].

The treatment of choice for extracardiac rhabdomyomas is surgical excision. Despite of the application of treatment regiments, relapse is possible. The risk of recurrence can equal even 42% [5]. This is probably due to the non-radical resection of the tumor and its multifocal development, which occurs in 14–26% of cases. Extracardiac rhabdomyomas do not have a tendency to malignancy [8].
**Case Report**

**Rhabdomyoma** is usually characterized with a slow growth and the presence of a painless mass in the head and neck. Signs and symptoms depend on the tumor location. The main symptoms described in literature include: upper airway obstruction, hearing loss, hemoptysis, dysphagia, hoarseness and odynophagia [9]. Furthermore, ARM in the pharyngeal space involves: the presence of a tumor in the submandibular triangle, asymmetry of the nasopharyngeal cavity and obstructive sleep apnea. Therefore, **rhabdomyoma** should be considered in differential diagnosis of tumors in this region along with: multiformal adenoma, periodontal disease, neuroblastoma, sarcomas and squamous cell carcinomas [8].

Histologically, these tumors are characterized by layers of densely packed round or oval cells with abundant, eosinophilic, granular cytoplasm. Cell nuclei are small and occupy a central or peripheral place in the cell. There are no signs of mitosis and necrosis [8]. Moreover, they have specific immunohistochemical properties, such as positive cytoplasmic for muscle-specific action, desmin and myo-D1 [9].

Preoperative diagnosis of extracardiac **rhabdomyoma** is challenging. Due to the excellent high-resolution determination of soft tissue, MR is the imaging method of choice for the diagnosis of pharyngeal space tumors. In magnetic resonance imaging, ARM is slightly hyperintensive or isointensive to muscles in T1- and T2-dependent images. Signal intensity is uniform within the lesion, but mild contrast enhancements may also appear [9, 10].

The presented case report shows that ARM has a high tendency to occur in the head and neck, and it can mimic other cancerous processes found in this location. In this case, both the clinical and radiological picture of the tumor did not allow for an unequivocal preoperative diagnosis. Management of the patient is an example of a misdiagnosis, which we may encounter in pharyngeal tumors, since it is the enlargement of the palatine tonsil and difficulty in swallowing that most often suggest pathology of the tonsil, especially since the lesions are unilateral. However, the MR examination is always the decisive factor; in the vast majority of cases, it illustrates the location of pathology and facilitates making decisions regarding treatment.

### References


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**Competing interests:** The authors declare that they have no competing interests.

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