CASE REPORT

Retrochrycoid Intermediate Extracardiac Rhabdomyoma: A Rare Cause of Dysphagia

Ana Nobrega Pinto¹, Eurico Monteiro², Eduardo Breda³, Eduardo Ferreira⁴

Received on: 22 December 2019; Accepted on: 26 April 2023; Published on: xx xx xxxx

Abstract

Introduction: Rhabdomyomas are rare benign tumors of the skeletal muscle. Regarding location, they can be subdivided into cardiac and extracardiac subtypes. Considering histological features, they can be classified as fetal, juvenile (intermediate), and adult.

Case description: A 25-year-old male patient presented with a 3-month progressive dysphagia. Physical examination showed a smooth submucosal bulging of the right retrochrycoid region with anterior displacement of the ipsilateral larynx. Imaging studies [computed tomography (CT) scan and magnetic resonance imaging (MRI)] corroborated the findings in indirect laryngoscopy showing a well-defined nodular formation centered on the right retrochrycoid area that caused the anteromedial displacement of the right hemilarynx. The patient underwent transoral CO₂ laser biopsy and the final histopathologic diagnosis was extracardiac juvenile rhabdomyoma. A transoral laser-assisted resection of the tumor was performed.

Conclusion: Although muscle tumors of the hypopharynx are uncommon, rhabdomyoma should be considered in the differential diagnosis of submucosal masses in this region. Complete surgical excision is the treatment of choice.

Keywords: Carbon dioxide lasers, Case report, Histology, Hypopharynx, Rhabdomyoma.

Otorhinolaryngology Clinics: An International Journal (2024): 10.5005/jp-journals-10003-1498

Introduction

Rhabdomyomas are rare benign tumors of skeletal muscle, accounting for less than 2% of all primary striated muscle tumors.¹ Based on location and histology, rhabdomyoma can be classified into several subtypes, as shown in Table 1.²³

Cardiac rhabdomyomas are most frequently multiple lesions that occur in children and are considered hamartomatous lesions.¹⁴

Fetal rhabdomyoma presents as an undefined mass or polypoid growth, more common in males with a wide age range. They are constituted of immature, spindle-shaped skeletal muscle elements in several stages of differentiation, with few mature cells.⁵

Adult extracardiac rhabdomyomas generally present as well-defined solitary lesions in middle-aged males and can be multifocal in 3–26% of cases.¹²

Histologically, intermediate rhabdomyomas fall somewhere between fetal and adult rhabdomyoma. This variant is 2 times more common in males tends to occur in adolescents or young adults and involves mucosal sites and soft tissues of the face.²³⁶

Most extracardiac rhabdomyomas occur in the head and neck region and are of adult type.⁵⁷

Case Description

A 25-year-old male patient presented with a 3-month progressive dysphagia associated with a foreign body sensation in the throat. He denied other complaints, such as dyspnea, dysphonia, or pain.

Table 1: Rhabdomyomas classification²³

<table>
<thead>
<tr>
<th>Site</th>
<th>Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac</td>
<td>Fetal</td>
</tr>
<tr>
<td>Extracardiac</td>
<td>Juvenile or Intermediate</td>
</tr>
<tr>
<td></td>
<td>Adult</td>
</tr>
</tbody>
</table>

The patient had already undergone two biopsies at another center and both results were inconclusive. He had no other relevant past medical or surgical history.

Physical examination showed a smooth submucosal bulging of the right retrochrycoid region with anterior displacement of the ipsilateral larynx. Vocal cords were moving normally and no other relevant alterations were identified.

Computed tomography (CT) scan and magnetic resonance imaging (MRI) were carried out and both showed a nonenhancing well-defined nodular formation centered on the right retrochrycoid region measuring 2.5 × 1.3 cm (transverse and anterior–posterior...
Retrochorycoid Intermediate Extracardiac Rhabdomyoma

Although clinical and imagiological findings suggested a benign lesion, malignancy could not be excluded, so the patient underwent a transoral CO$_2$ laser biopsy under general anesthesia for histological characterization.

Microscopically, the tissue sample was constituted by large round and polygonal eosinophilic cells, with predominantly centrally placed, nuclei that were separated by small undifferentiated spindle cells (Fig. 3). Cross-striations were identified and there was no mitotic activity or nuclei atypia. Immunohistochemically, cells strongly expressed desmin (Fig. 4). They were also negative for smooth muscle-specific actin (Fig. 4), CD34, and cytokeratin MNF116.

The final histopathologic diagnosis was intermediate rhabdomyoma.

Subsequently, a transoral laser-assisted resection of the tumor was carried out (Fig. 5). There were no complications to the surgical procedure and the patient recovered fully, without any complaints of dysphagia or hoarseness.

**Discussion**

The most frequently reported location of rhabdomyoma in the larynx is the glottis, with very few cases described in the hypopharynx. To our knowledge, four cases of rhabdomyomas have been described in this location (Table 2). Preoperative diagnosis of extracardiac rhabdomyomas can be challenging. The tumor usually presents as a submucosal mass with a smooth surface and may look like a cyst. Radiographically it presents as a homogenous lesion, isointense or slightly hyperintense to muscle on T1- as well as T2-weighted MRI and slightly hyperdense on CT. Histological examination is essential for a definite diagnosis. Care should be taken when performing a biopsy of these submucosal lesions to ensure proper collection of affected tissue and not only normal mucosal lining. Also, since these tumors are
Figs 3A and B: Microscopic view of tissue specimen: Eosinophilic round or polygonal cells with a granular or vacuolated cytoplasm separated by spindle cells [hematoxylin and eosin; magnification: (A) 10x; (B) 40x]

Figs 4A and B: (A) Immunohistochemistry for desmin and for smooth muscle-specific actin; (B) Rhabdomyoblasts show diffuse cytoplasmic reactivity to desmin and no cytoplasmic reactivity to smooth muscle-specific actin. Cross-striations can be observed (magnification: 40x)

Figs 5A and B: (A) Intraoperative endoscopic image of the bulging submucosal lesion on the right retrochrycoid area; (B) Endoscopic view after surgical excision
Table 2: Reported cases of hypopharynx rhabdomyoma

<table>
<thead>
<tr>
<th>S. No.</th>
<th>Author</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Koutsimpelas D et al.⁴</td>
<td>Left hypopharynx</td>
</tr>
<tr>
<td>2</td>
<td>Winther⁸</td>
<td>Left vallecular</td>
</tr>
<tr>
<td>3</td>
<td>Lamoral et al.⁹</td>
<td>Posterior wall of the left pyriform sinus (extension to midline)</td>
</tr>
<tr>
<td>4</td>
<td>Koudounarakis et al.¹⁰</td>
<td>Right pyriform sinus</td>
</tr>
</tbody>
</table>

Local recurrences are usually related to incomplete primary resection and its frequency varies from 10 to 42%.⁴ Due to the slow growth nature of these tumors the recurrent lesions may only be identified many years after initial resection and so a close follow-up of these patients is advisable.⁴ There are no reported cases of malignant transformation.⁴

References