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CASE REPORT

Rhabdomyoma of the Parapharyngeal Space Presenting with Dysphagia

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Abstract Rhabdomyoma is an exceedingly rare soft tissue benign tumor of skeletal muscle origin classified into cardiac and extracardiac types based on location. Extracardiac rhabdomyoma is further classified into adult, genital, and fetal type depending on the degree of differentiation. Adult rhabdomyomas are rare, but morphologically characteristic, benign mesenchymal tumors with mature skeletal muscle differentiation that in 90% of cases arise in the head and neck region, mainly in the mucosa of the oropharynx, nasopharynx, and larynx, from the branchial musculature of third and fourth branchial arches. Most patients are between 40 and 70 years old, with a mean age of 60 years with a male predominance. Usually presenting symptoms include upper airway obstruction, Eustachian tube dysfunction, and mucosal or neck mass, but rarely does it arise as pure dysphagia. This article presents a case of parapharyngeal rhabdomyoma presenting with only progressive dysphagia.

Keywords Rhabdomyoma · Parapharyngeal space · Benign mesenchymal tumor · Pharynx neoplasm · Dysphagia · Deglutition · Deglutition disorders

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Rhabdomyoma is an exceedingly rare soft tissue benign tumor of skeletal muscle origin. Although as a general rule most benign soft tissue tumors outnumber their malignant counterparts, this does not hold true for striated muscle tumors as rhabdomyomas account for only 2% of skeletal muscle tumors [1]. Rhabdomyoma is a benign mesenchymal tumor with skeletal muscle differentiation that is classified into cardiac and extracardiac types based on location [2]. Extracardiac rhabdomyoma is further classified into adult, genital, and fetal type depending on the degree of differentiation, although some overlap exists [3]. Unlike cardiac rhabdomyoma, there is no association with tuberous sclerosis [4]. Adult rhabdomyomas are rare, but morphologically characteristic, benign mesenchymal tumors with mature skeletal muscle differentiation which in 90% of cases arise in the head and neck region, mainly in the mucosa of the oropharynx, nasopharynx, and larynx, from the branchial musculature of third and fourth branchial arches. Most patients are between 40 and 70 years old, with a mean age of 60 years with a male predominance [5]. Usually presenting symptoms include upper airway obstruction, Eustachian tube dysfunction, and mucosal or neck mass but it rarely arises as pure dysphagia. Adult-type rhabdomyomas are often solitary but may be multinodular or rarely multifocal.

Case Report

A 56-year-old male patient was referred to the Otolaryngology Head and Neck Surgery Department of the National Cancer Institute of Rome with a three-year history of a slowly growing asymptomatic oropharyngeal mass. Two months before admission the patient developed progressive dysphagia initially only for liquid, then for solid food. Physical examination showed a bulky lesion covered with

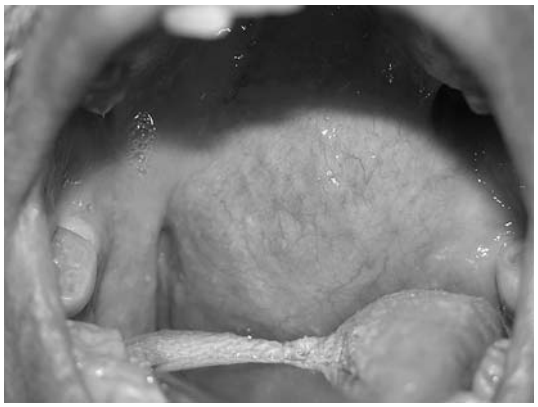

Fig. 1 Oropharyngeal bulky lesion

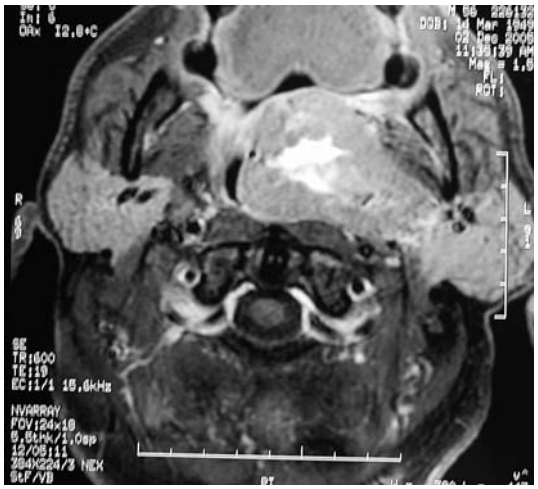

Fig. 2 MRI scan shows a 7.5 × 5 × 3.5-cm large mass in the left prestyloid parapharyngeal space (axial image)

normal mucosa involving three quarters of the oropharynx on the left side (Fig. 1). Nasopharyngoscopic examination showed the lesion occupying the left side of nasopharynx, the left pyriform sinus, the left aryepiglottic fold, and the laryngeal side of the epiglottis. The vocal fold moved well.

Magnetic resonance imaging (MRI) was acquired on a 0.5-T superconductive system using the SE technique and T1, T2, and fat-suppressed sequences before and after contrast medium (gadolinium-DTPA) intravenous infusion. The MRI scan showed a 7.5 × 5 × 3.5-cm large mass in the left prestyloid parapharyngeal space. The carotid artery was patent and there were no signs of infiltration of the surrounding tissue (Figs. 2–4).

The patient underwent a tracheotomy under local anesthesia, because it was impossible to do orotracheal or nasotracheal intubation, a total parotidectomy via the transcervical approach, and resection of the lesion without mandibulotomy. The whole tumor was sent to pathology as one gross specimen. Macroscopically the mass had a maximum diameter of 7.6 cm, was circumscribed, and, on

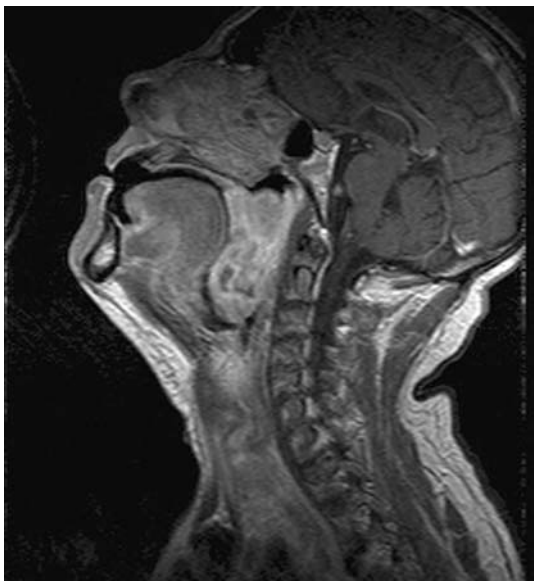

Fig. 3 Coronal MRI image

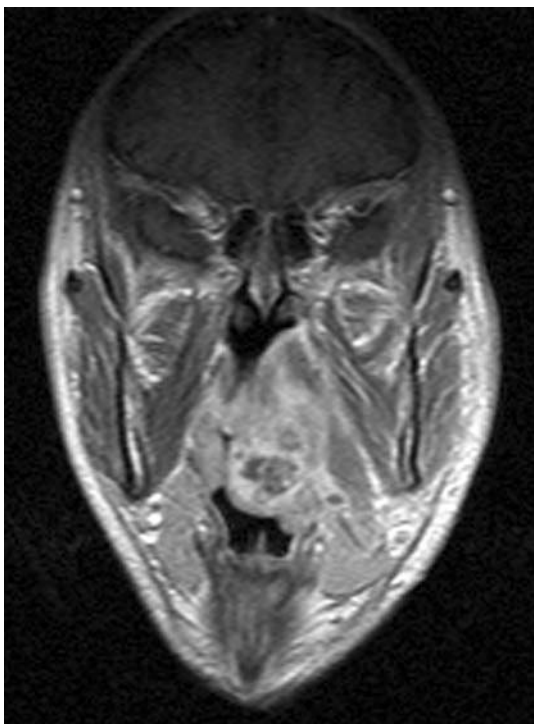

Fig. 4 Sagittal MRI image

cut surface, was deep tan to red-brown, soft, and lobulated. Microscopically, it appeared as a well-defined but unencapsulated mass composed of lobules of closely packed, uniform, large, polygonal cells in a scant stroma. The cells had abundant eosinophilic, granular cytoplasm with well-defined borders, and vesicular, small, round, centrally located nuclei, sometimes with prominent nucleoli. Cross striations and intracytoplasmic rodlike inclusions were frequently seen. There was no mitotic activity or nuclear

atypia. The glycogen-rich cytoplasm was periodic acid-Schiff (PAS) positive. Masson trichrome stain highlighted the cytoplasm striation as well as the rodlike inclusion.

The postoperative course was uneventful and the patient was discharged six days after intervention.

Discussion

Extracardiac rhabdomyomas are very rare tumors comprising less than 2% of the neoplasm of the striated origin. This entity was first described in 1897 by Pendl, who presented a case of fetal type [6], and fewer than 100 cases have been reported in the literature. Extracardiac rhabdomyomas prefer the head and neck region, arising from the musculature of the third and fourth branchial arches. The gold standard treatment is surgery. It can recur, but it never turns malignant. Recurrence may be as high as 42% [4] and there has been one case with three recurrences in a 35-year period [7]. This is probably due to incomplete excision or multifocality which is seen in 14–26% of the cases [7–9].

The signs and symptoms at time of presentation depend on where the tumor is growing. Some of the symptoms from the reported cases in the literature are hearing loss, mass in the submandibular triangle, swelling in the nasopharynx, hemoptosis, and obstructive sleep apnea but rarely pure dysphagia [4, 5, 10]. Therefore, rhabdomyoma should be considered in a differential diagnosis in this region, such as alveolar soft part sarcoma, granular cell tumor, hibernoma, rhabdomyosarcoma and other sarcoma of head and neck, oncocytoma, paraganglioma, and tumor of salivary glands.

On MRI the rhabdomyoma appeared isointense with the muscle on T1-weighted images and slightly hyperintense in T2 and in the fat-suppressed sequences; the signal intensity was homogeneous after Gd-DTPA infusion and the lesion showed moderate enhancement. The signal intensity of the rhabdomyoma was different than pleomorphic adenomas of the minor salivary gland and of the deep portion of the parotid gland that show a low signal on T1-weighted sequences and high intensity on T2 and fat-suppressed sequences with a marked enhancement after Gd-DTPA infusion. The signal of these tumors can be heterogeneous for the presence of high-signal-intensity areas on T1 sequences in relation to proteinaceous fluid; on T2 sequences cystic zones appear as an area of the high intensity without enhancement after Gd-DTPA infusion. The signal intensity of the carcinoma tends to be similar to that of muscle on T1 sequences and slight hyperintense on T2 sequences with homogeneous enhancement after Gd-DTPA infusion as a rhabdomyoma. However, the carcinomas usually appear as an infiltrative mass, contrary to rhabdomyoma which has more regular margins [10].

The immunohistochemical features of rhabdomyoma are identical to those of normal skeletal muscle cells because it shows cytoplasmic positivity for MSA (muscle-specific actin), desmin, and MyoD1. The histologic appearance of adult rhabdomyoma with its large, polygonal skeletal muscle cells is very distinctive [11, 12].

Conclusion

Although rare, adult extracardiac rhabdomyoma is most commonly found in the head and neck region and should be considered in the differential diagnosis of masses in this region, as well as the cause of progressive dysphagia.

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