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Case Report

Adult rhabdomyoma of larynx: a case report

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Abstract: Rhabdomyoma is a rare benign tumor derived from skeletal muscles. Adult rhabdomyoma in larynx is extremely uncommon and has rarely been reported. Here, we reported a case of adult rhabdomyoma of a 39-year-old woman. Initially the patient had a diagnosis of granular cell tumor on a fine needle aspiration (FNA). Laryngoscopy revealed a large submucosal mass was noted in the left subglottic larynx. The contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) of the neck revealed a well-demarcated diffusely hyperattenuating submucosal mass on the left subglottic larynx. Pathology report showed adult rhabdomyoma, and curative treatment was obtained by surgery. There was no obvious recurrence.

Keywords: Rhabdomyoma, larynx, surgery

Introduction

Rhabdomyoma (RM) is a rare benign neoplasm of striated muscle, which can be divided into cardiac and extracardiac forms. Most rhabdomyomas arise from cardiac muscle. Cardiac rhabdomyoma occurs almost exclusively in the pediatric age group and may be associated with tuberous sclerosis, neurofibromatosis, and sebaceous adenomas [1]. Of the extracardiac ones, there are three subtypes: adult, fetal, and genital. The adult type has a predilection for the head and neck of elderly males. These tumors originate from the branchial musculature of the third and fourth branchial arches [2]. Seventy seven percent of all extracardiac rhabdomyomas occur in the head and neck and 14% occur in the genital region [3]. Adult rhabdomyoma is the most common subtype of rhabdomyoma even though it remains relatively rare. In this paper, we reported a rare case of a rhabdomyoma originating from left subglottic larynx.

Case report

The 39-year-old woman complained of progressive hoarseness during a 2-year period. The patient had a diagnosis of granular cell tumor on an fine needle aspiration (FNA) from an outside institution. As per the hospital protocol, her outside pathology slides were requested for review. We received FNA slides consisting of papanicolaou stained smears, H and E stained cytospin preparations, and a manually stained cell block slide with limited material.

On indirect examination of the larynx, a large submucosal mass was noted in the left subglottic larynx (**Figure 1**). The contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) of the neck revealed a well-demarcated diffusely hyperattenuating submucosal mass on the left subglottic larynx (**Figure 2**). The patient was taken to the operating room where the mass was resected, and immunohistochemical (IHC) stains performed on the histologic sections revealed the lesion to be diffusely positive for muscle-specific actin, desmin and myoglobin (**Figure 3**). Markers for epithelium membrane tumors and S-100 protein for neuroectoderm tumors were all negative, which supported the diagnosis of adult rhabdomyoma.

At a careful revision of the slides, it revealed a cellular aspirate composed of large cells with well-defined, abundant, dense eosinophilic cytoplasm and mostly peripherally located nuclei (**Figure 4**). Nuclei were round, vesicular, cen-

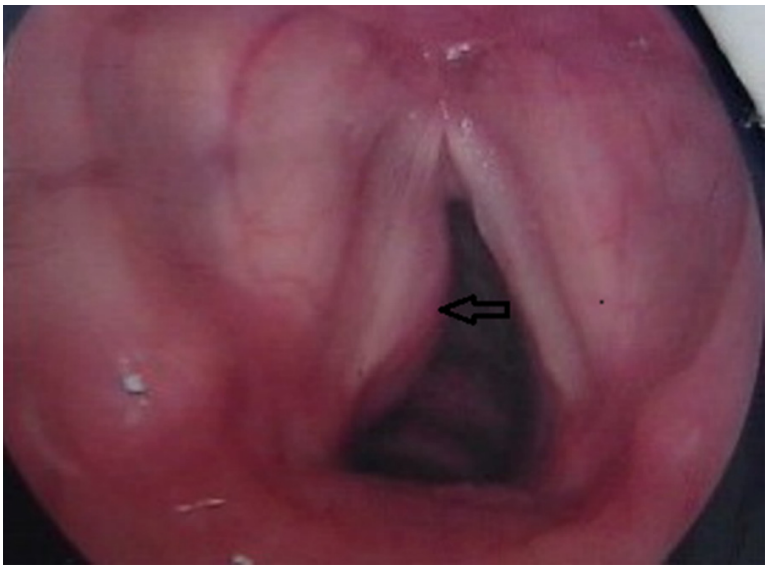

Figure 1. Laryngoscopy image showing submucosal mass of the left subglottic larynx.

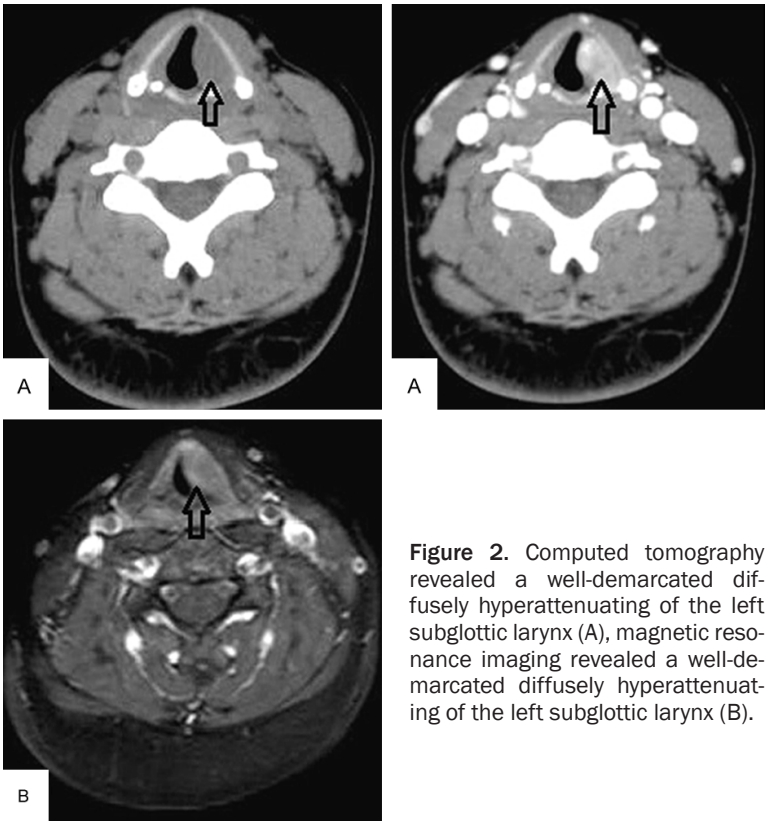

Figure 2. Computed tomography revealed a well-demarcated diffusely hyperattenuating of the left subglottic larynx (A), magnetic resonance imaging revealed a well-demarcated diffusely hyperattenuating of the left subglottic larynx (B).

trally located, with prominent nucleoli. Intracytoplasmic and intranuclear hyaline inclusions were seen. Cross-striations, not been noted before, were present in few cells. Mitotic activity and necrosis were absent. The patient recovered well. But, her voice was still hoarseness postoperatively. She is free of recurrence at 15 months of follow-up.

Discussion

Rhabdomyoma is an exceedingly rare soft tissue benign tumor derived from skeletal muscle. The pathogenesis is unclear. As a general rule, benign soft tissue tumors occur more frequently than their malignant counterparts, but this does not hold true for striated muscle tumors because rhabdomyomas account for only 2% of skeletal muscle tumors [2, 4, 5].

Certainly the adult-type RM of the larynx is an extremely rare benign tumor of myoblastic origin. A comprehensive list of adult-type laryngeal RMs reported in the literature is summarized in **Table 1**. In the past, RM was confused with other similar tumors, especially with granular cell tumors. This confusion is due to granular eosinophilic cytoplasm that is common to both diagnoses, although they have a completely different histogenesis. Another important observation of the histologic point of view is that the RM should not be confused with rhabdomyosarcoma, which has a completely different prognosis. The histologic diagnosis should be no problem if adequately prepared slides are available, but as these tumors are very rare and many pathologists are not familiar with this type of tumor, there are reports of substantial difficulties to diagnose them on the first try. Genital rhabdomyoma is seen most frequently in the vagina or vulva of young and middle-aged women. Clonal structural chromosomal abnormalities including

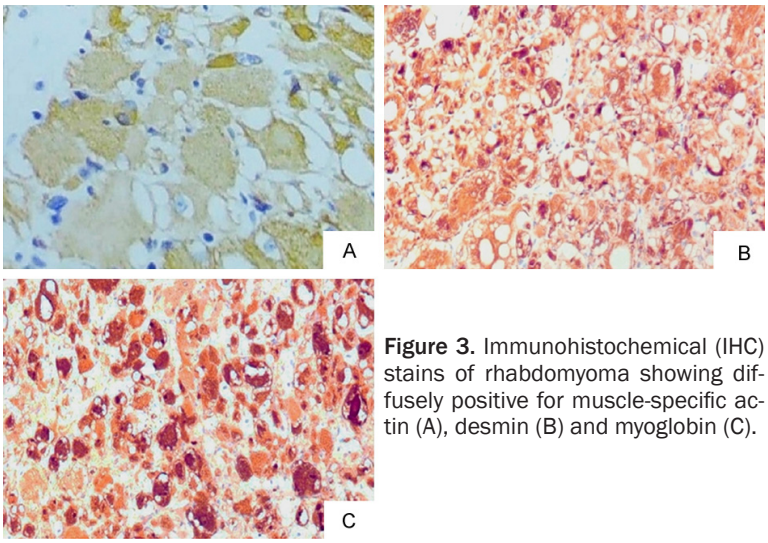

Figure 3. Immunohistochemical (IHC) stains of rhabdomyoma showing diffusely positive for muscle-specific actin (A), desmin (B) and myoglobin (C).

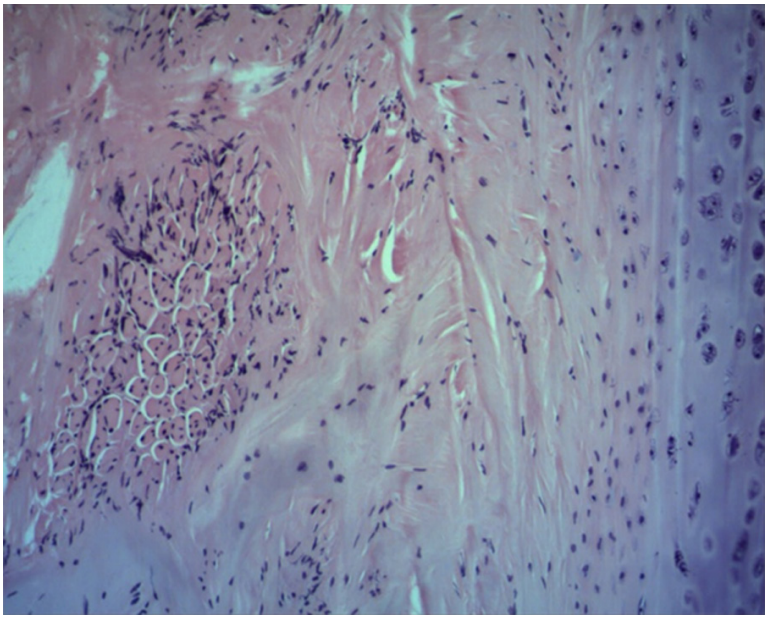

Figure 4. Histology of rhabdomyoma showing closely packed polygonal cells with deeply eosinophilic vacuolated cytoplasm.

reciprocal translocation between chromosomes 15 and 17 (in the majority) and abnormalities of the long arm of chromosome 10 have been demonstrated in these tumors, indicating that they are neoplasms rather than hamartomas [3, 6-8]. Given that these tumors are often found in the head and neck area, they lend themselves to fine needle aspiration (FNA).

We presented a case of laryngeal RM in a patient whose diagnosis was not initially set. Although the clinical and pathologic findings should be taken into consideration, we emphasized that IHC plays an important role in the differential diagnosis. Symptoms of laryngeal RM donot differ from those caused by other benign tumors of the larynx, but one should keep in mind the possibility of local airway obstruction when the patient complains of breathing difficulties. The tumor is usually described as a submucosal mass with a smooth surface that can resemble the appearance of a cyst. Laryngoscopy and imaging exams help locate and define the limits of the tumor, but the definitive diagnosis will always depend on the histopathologic studies. Adult rhabdomyomas are positive for desmin, myoglobin, and muscle-specific actin, and negative for S100 and CD68. Granular cell tumors tend to be diffusely positive for S100 and CD68. It is important to note that adult rhabdomyomas may show focal positivity to S100, so more than one stain should be performed when the differential diagnosis of adult rhabdomyoma and granular cell tumor is under consideration [9-11].

The clinical outcome of laryngeal RM adult type is benign and no evidence of metastasis was reported in the literature. Treatment should always be surgical removal, via either endoscopic or external approach, but always preserving the adjacent structures such as the vocal folds and the swallowing apparatus. Recurrence can happen but usually is due to incomplete excision of the tumor. When total excision cannot be accomplished, reoperation or narrow follow-up is indicated to prevent advanced revision surgeries. In this presented case, complete excision was done, and there was no obvious recurrence.

In conclusion, the rhabdomyoma is distinctly rare. Our patient was a rare case of adult rhab-

Table 1. Adult rhabdomyoma of the larynx and their characteristics

Author	Sex	Age (year)	Location	Chief complaint
Clime ARW (1963)	Male	48	Right vocal cord	Hoarseness for 3 months
Battifora HA (1969)	Male	55	Glottis	Hoarseness for 3 years
Bianchi C (1975)	Female	52	Right false vocal cord	Disphonia for 3 years
Bagby RA (1976)	Male	55	Right false vocal cord	NA
Ebbesen (1976)	Female	64	Right ventricle	Hoarseness, foreign-body sensation for 6 months
Winther LK (1976)	Male	39	Left vocal cord	Hoarseness for 3 years
Boedts D (1979)	Female	76	Left vocal cord	Hoarseness for 2 months
Kleinsasser O (1979)	Male	16	Glottis	Acute airway obstruction sudden onset
Modlin B (1982)	Male	52	Left vocal cord	Hoarseness for 6 months
Heliwell TR (1988)	Male	66	Right vocal cord	Hoarseness for 8 years
Hamper K (1989)	Female	51	Aryteniod	Dyspnoea, dysphagia
Selme V (1994)	Female	31	Vocal cord	Hoarseness
Roberts DN (1994)	Female	56	Interarytenoid	Dramatically worsening dysponea and mild dysphagia for 6 weeks
Zbären P (1995)	Male	64	Left aryepiglottic fold	Asymptomatic submandibular mass
Johansen EC (1995)	Male	51	Left ventricle	Hoarseness, snoring for 1year
LaBagnara J Jr (1999)	Female	69	Vocal cord	Hoarseness for 5 years
Orrit JM (2000)	Male	66	Aryteniod	Hoarseness and dysphagia for 4 months
Liang GS (2000)	Male	39	Left-sided paraglottic space larynx	Dysphagia and weight loss for 5 months
Brys AK (2005)	Male	79	Right false vocal cord	Hoarseness for 5 years
Liess DB (2005)	Male	69	Epiglottis	Asymptomatic submandibular mass
Jensen K (2006)	Male	66	Right arytenoid	Dysphagia and hoarseness for 3 years (sudden dyspnea)
Formigon GG (2006)	Male	35	Extrinsic laryngeal muscles	Cervical bulging
Koutsimpelas D (2008)	Female	72	Left aryepiglottic fold	Globulus and hoarseness for 1 year
Farboud A (2009)	Male	76	Arytenoid	Hoarsness, dysphagia and sleep-apnoea
Friedman AD (2012)	NA	NA	Glottis	Dysphonia
de Trey L A (2013)	Male	55	Left paraglottic space	Hoarseness and slight dysphagia for 3 months
Cain RB (2013)	Female	67	Supraglottis	Hoarseness and progressive dyspnea
Pinho MM (2013)	Male	35	Supraglottic	Progressive hoarseness for 1 year
Vijendra Shenoy S (2013)	Male	50	Right aryepiglottic fold	Change in voice for 2 years
Carta F (2016)	Male	75	Right false vocal cord	Progressive dysphonia for 4 years
Altissimi G (2017)	Female	85	Right arytenoid	Sudden breathing difculties, dysphagia for 10 days
Balakumar R (2018)	Female	51	Supraglottic	Birt-Hogg-Dubé syndrome for 3 years
Present case	Female	39	Left subglottic	Hoarseness for 2 year

domyoma located in left subglottic larynx. Initially the patient had a diagnosis of granular cell tumor on an FNA. Pathology report showed adult rhabdomyoma, and curative treatment was obtained by surgery. There was no obvious recurrence. Although rhabdomyoma of larynx is a rare disease, we should keep in mind this differential diagnosis when we meet the patient. Be sure to take photo documentation routinely before punch biopsy or excision even though it looks like benign lesion. Surgical excision is an imperative treatment for rhabdomyoma. Recurrence rate is extremely low when complete excision is done.

Disclosure of conflict of interest

None.

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