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CASE REPORT

Rhabdomyoma of larynx – A diagnaostic dilemma

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KEYWORDS

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Abstract *Introduction:* Rhabdomyomas of the head and neck are unusual. Extracardiac rhabdomyoma is a rare benign tumor of striated muscle origin that is found in adults and children. We report a case of rhabdomyoma of the larynx, which presented a diagnostic dilemma.

Case report: We report an uncommon case of a 50-year-old man, who presented with change in voice for two years. Endoscopic examination of the larynx revealed a smooth mass covered by intact mucosa involving the right aryepiglottic fold and a false vocal cord with restricted right vocal cord mobility. Biopsy from the lesion revealed a diagnosis of squamous cell carcinoma. The patient underwent total laryngectomy. To our surprise, histologic examination and immunohistochemical diagnosis proved it to be rhabdomyoma of the adult type with no evidence of malignancy.

Discussion: Most extracardiac rhabdomyomas arise from the pharyngeal constrictor muscles, the floor of the mouth, or the base of the tongue. Rarely, rhabdomyomas may arise from the larynx. Histologic and immunohistochemical analysis is essential for diagnosis. Treatment of rhabdomyoma consists primarily of complete excision with preservation of the surrounding tissues.

Conclusion: Our case demonstrates various differential diagnoses possible in laryngeal neoplasms. It also shows that immunocytochemistry plays an important role in differential diagnosis.

1. Introduction

Rhabdomyomas are benign tumors of skeletal muscle derivation. They are most commonly found in the heart as hamartomas associated with the condition of tuberous sclerosis. Most extracardiac rhabdomyomas occur in the head and neck and arise from the pharyngeal constrictor muscles, the floor of the mouth, or the base of the tongue. Rarely, rhabdomyomas may arise from the larynx. We report a rare case of laryngeal rhabdomyoma, which presented a diagnostic dilemma.

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2. Case report

A 50 year old male presented to our outpatient department, with change in voice for the past two years which was non progressive. He gave no history of voice abuse, fatigue, or any diurnal variation. There was no history of breathing difficulty, dysphagia or throat pain. He had no significant weight loss or loss of appetite, and was not suffering from any chronic illnesses. Laryngeal endoscopy revealed a mass covered by intact mucosa involving the right aryepiglottic fold, false cord and bulging into the medial wall of the right pyriform fossa. Vocal cord mobility was restricted on the right side. (Fig. 1) There were no palpable neck nodes. His routine blood investigations and chest X-ray were found to be normal. A contrast enhanced computed tomography of the neck was done, which showed a well defined homogenous and intensely enhancing mass in the right aryepiglottic fold bulging into the right pyriform fossa. There is no erosion of thyroid cartilage The lesion was limited within the thyroid lamina laterally, up to the lower border of the epiglottis superiorly, and was seen extending up to the level of the glottis involving the right true cord inferiorly (Fig. 2). We planned for a direct laryngoscopy and hypopharyngoscopy with biopsy of the lesion under general anaesthesia. Intubation for general anaesthesia proved difficult, so we went ahead with a tracheostomy. Direct laryngoscopy revealed a large mass covered by intact mucosa on the right arytenoid, right aryepiglottic fold and medial wall of the right pyriform fossa, pushing the larynx to the left. Bilateral vocal cords were visualized to be normal. Biopsy was taken from the mass and sent for histopathology, which provided a tissue diagnosis of squamous cell carcinoma with micro-invasion. It was subsequently decided that total laryngectomy was the best possible means of treatment.

The patient underwent total laryngectomy with partial pharyngectomy, bilateral neck dissection and primary reconstruction under general anaesthesia. Intraoperative findings included a soft submucosal growth involving the right aryepiglottic fold, right false cord, and medial wall of the right pyriform fossa. There was no induration or ulceration of the lesion. Histopathological examination of the specimen revealed a circumscribed tumour in the paraglottic space, with sheet like proliferation of tightly packed large polygonal to round cells with abundant deeply eosinophilic granular cytoplasm and centrally and peripherally placed nuclei. Some of the cells showed vacuolated cytoplasm due to glycogen which stained positive with PAS stain. PTAH stain showed positive rod like inclusion bodies (Fig. 3). Immunohistochemistry was done, and was found to be negative for chromogranin, synaptophysin, CK and S-100, and focal positivity for vimentin and SMA, and strong positivity in many cells for desmin, giving a final diagnosis of rhabdomyoma of the adult type (Fig. 4). We could not perform immunohistochemistry for myogenin and MyoD1, because of non-availability in our institution. The

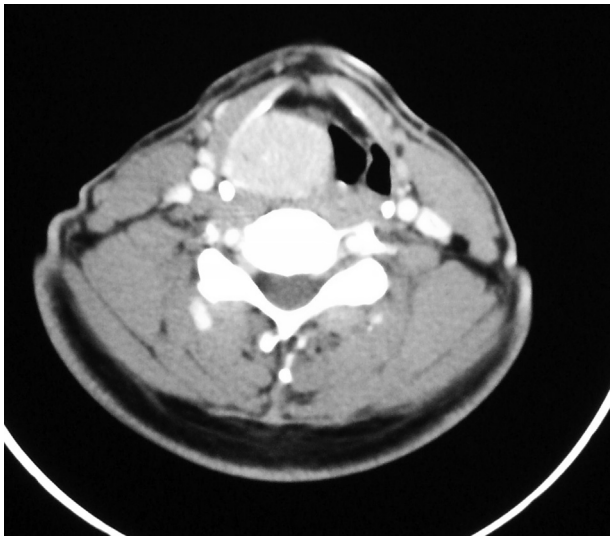

Figure 2 CT contrast showing a well defined homogenous and intensely enhancing mass in the right aryepiglottic fold bulging into right pyriform fossa. There is no erosion of thyroid cartilage.

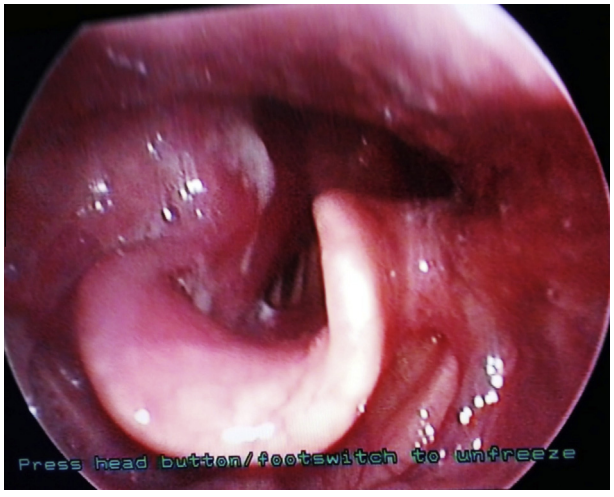

Figure 1 Laryngoscopy showing a large, mucosa covered smooth bulge in the region of the right arytenoid, right aryepiglottic fold and medial wall of right pyriform fossa, pushing the larynx to the left.

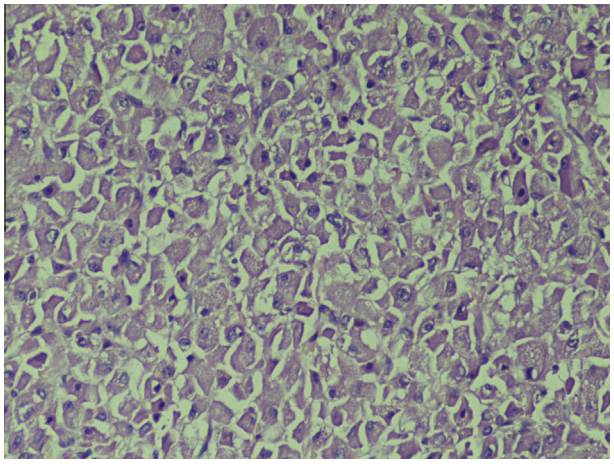

Figure 3 Histopathological examination showing large round to polygonal cells with eosinophilic granular cytoplasm and vesicular nuclei. Some of the cells show vacuolated cytoplasm.

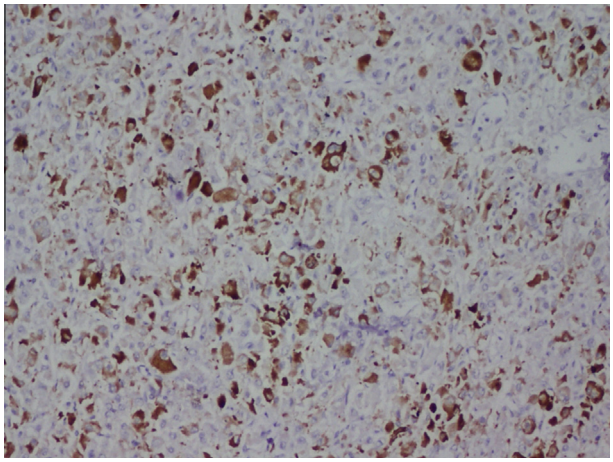

Figure 4 Immunohistochemistry slides showing strong positivity seen in many cells with desmin.

initial biopsy slides, which were reported as squamous cell carcinoma, were reviewed and deeper sections were studied. It was showing a severe degree of pseudoepitheliomatous hyperplasia with inflammatory reaction, which was misinterpreted as microinvasion. The patient recuperated well from the surgery. A tracheo-esophageal prosthesis insertion was done as a second stage procedure for voice restoration. The patient has come for regular follow up for the past 6 months, and remains disease free to date.

3. Discussion

Extracardiac rhabdomyoma is a rare benign tumor of striated muscle origin that is found in adults and children. It is considered distinct from cardiac rhabdomyoma, which is believed to be a hamartoma and not a true neoplasm and which is usually diagnosed in children and associated with tuberous sclerosis. The term rhabdomyoma was first used by Zenker (1864) and the first laryngeal rhabdomyoma was described by Imperatori (1933).[1] The largest case series of extracardiac rhabdomyomas reviewed is that by Helliwell et al. in 1988, with 115 cases, of which 70% were located in the head and neck region and 15 in the larynx.[2] In 1995, Johansen et al. published a series of 23 cases of laryngeal rhabdomyoma.[3] Of these, 15 were of the adult type, 4 fetal cellular, and 4 fetal myxoid. Some of the cases of adult rhabdomyomas of the upper respiratory tract reported in the literature[1–13] are shown in Table 1.

Adult rhabdomyoma occurs more frequently in men than women (4:1). The reported ages of affected patients have ranged from 16 to 76 years (mean: 52).[4] Rhabdomyomas are slow growing tumours. The most common symptoms of laryngeal rhabdomyoma are hoarseness, a foreign-body sensation, and dysphagia. The duration of symptoms is usually long, typically years. The main presenting symptom of our case was a change in voice for two years. Most reported cases of extracardiac rhabdomyoma have involved solitary masses. Rhabdomyomas are generally benign, never metastasize, and rarely recur.[4]

Of muscle tumors generally, rhabdomyomas actually occur less commonly than rhabdomyosarcomas, representing less than 2% of all striated muscle tumors.[5] The most frequent site of occurrence of these benign muscle tumors is the myocardium. Extracardiac rhabdomyomas are found frequently in the head and neck because they arise embryologically from the unsegmented mesoderm of the branchial arches and not from myotomes like the rest of the skeletal muscles.[6]

Radiographically, adult rhabdomyomas appear as benign neoplasms. However, indistinct borders blending into adjacent muscle can appear as malignant lesions on CT. Magnetic resonance imaging may help to delineate the extent and location of tumor involvement. On T1- and T2-weighted images, rhabdomyomas are isointense or slightly hyperintense to muscle, and they homogeneously enhance without necrosis or hemorrhage.[7]

Grossly, the tumors appear as well defined, rounded, or coarsely lobulated masses. They range from 0.5 to 10 cm in greatest diameter. They may be either multinodular or form sessile or pedunculated polypoid submucosal masses. On cut section it has a finely granular, gray–yellow to red brown appearance. Histological examination is essential for

Table 1 Comparison of our case of laryngeal rhabdomyoma with other reported cases in the literature.

1.	Roberts et al.[1]	56	F	Inter arytenoid area	Excision	Desmin positivity, SMA negative
2.	Johansen et al.[3]	51	M	Left ventricle	Excision	Desmin, myoglobin, lactin positive
3.	Jensen et al.[4]	66	M	Vocal process of right arytenoid	Excision	Strongly positive for desmin
4.	LaBagnara et al.[5]	69	F	Right vocal cord	MLS + Biopsy	Not assessed
5.	Winther et al.[6]	54	M	Left hypo pharynx	Excision	Not assessed
6.		39	M	Left vocal cord	Excision	Not assessed
7.	Brys et al.[7]	79	M	Right vocal cord	External excision	Not assessed
8.	Viscasillas G et al.[8]	50	F	Right hemilarynx	Biopsy & transoral exeresis with CO2 laser	Not assessed
9.	Rosenman et al.[9]	78	F	Left glottis with Left VC palsy	Excision	Not assessed
10.	Modlin et al[10]	34	F	Right Supra glottis	Right lateral pharyngotomy Right supraglottic laryngectomy	Not assessed
11.	Hamper et al.[11]	51	F	Left supraglottic	Excision	Desmin,myoglobin,actin,S-100 positive
12.	Bagby et al.[12]	55	M	Right false cord	Excision	Not assessed
13.	Liang et al.[13]	39	M	Left supraglottic	Open biopsy & resection later	Not assessed
14.	Our case	50	M	Right paraglottic space	Total laryngectomy	Desmin positive, SMA and Vimentin focal positivity, S-100 negative

diagnosis. Three histological types are described, and each presents a different clinical picture. The adult type is most frequently found in the head and neck area of adults. These are slow-growing tumors with well-defined margins. The tumors are composed of dense sheets of closely packed large, round cells filled with eosinophilic cytoplasm. The cells have centrally or peripherally placed vesicular nuclei with prominent nucleoli. Many of the cells are vacuolated because of intracelluar glycogen dissolved during processing. Cross striation can be demonstrated with PTAH stain in most cases. The cytoplasm may be finely granular and should be differentiated from granular cell tumor. The cells of granular cell tumor tend to be less well defined and positive for S-100 protein. The fetal cellular type is extremely rare; it may occur in the head and neck of adults, but is most commonly found in young children less than 4 years of age. These tumors are poorly defined and consist of immature spindle-shaped skeletal muscle elements with few mature cells. The fetal myxoid or the genital type is found in the vagina and vulva of middle-aged women. These usually present as a polypoid mass filled with abundant loose, edematous and myxoid stroma.[5] Rhabdomyomas can be immunostained with antibodies to muscle specific actin, desmin, and myoglobin, which are markers of mature muscle cells.[7]

Histologically, our patient's neoplasm showed the classic features of adult rhabdomyoma: large, round cells with pink-staining, granular cytoplasm and vesicular nuclei. Rod like inclusions, and cross-striations were also found. Our case also showed positive staining with desmin.

Treatment of rhabdomyoma consists primarily of complete excision with preservation of the surrounding tissues. When possible, endoscopic excision is the preferred method of resection; however, for large or recurrent lesions, laryngotomy or laryngectomy may be necessary.[5] Recurrences are most commonly associated with incomplete excision and have been reported to occur in more than one-third of cases.[7]

We present this case for the rarity of laryngeal rhabdomyoma, to point out the pathological dilemma in diagnosis and to create awareness about immunohistochemistry to come to a definitive diagnosis. A benign tumour like rhabdomyoma should suspect when there is a long history with no lymph nodes. CT scan appearance of a well localized vascular mass not compatible with a case of usual squamous cell carcinoma should draw attention to revise the clinical diagnosis and look further into repeat biopsy or to discuss with the pathologist how to save the patient from laryngectomy. In a well circumscribed mass, covered with intact mucosa, the need of a deep biopsy after dividing the mucosa is essential to prevent misdiagnosis.

4. Conclusion

Laryngopharyngeal rhabdomyomas are uncommon tumours that are conventionally treated with surgical resection. Close collaboration with an experienced pathologist is necessary to establish the diagnosis with immunohistochemical evaluation to confirm the diagnosis. Surgery is the mainstay of treatment.

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