Case report

CO_2 Laser Resection of a Supraglottic Rhabdomyoma: Case Report and Review

J. Darcy O'Brien[2], James Belyea[1], Robert Hart[2], Jonathan Trites[2], S. Mark Taylor[2]
[1]*Class of 2011, Faculty of Medicine, Dalhousie University*
[2]*Division of Otolaryngology-Head and Neck Surgery, Department of Surgery, Dalhousie University, Halifax, Nova Scotia, Canada*

Abstract

Rhabdomyomas are rare benign tumours of striated muscle tissue that can be divided into cardiac and extracardiac types. Cardiac rhabdomyomas are associated with tuberous sclerosis, whereas extracardiac varieties are not associated with any particular syndrome. Approximately 70% of rhabdomyomas found outside the heart occur in the head and neck. Rhabdomyomas are typically solitary lesions, although multifocal lesions have been described; however, there have been no reports of malignant transformation to rhabdomyosarcomas. There have been 32 cases of laryngeal rhabdomyoma reported. Of these, 10 cases were reported in the supraglottic space. We present the 11th reported case of a supraglottic rhabdomyoma, and the first to be managed with laser resection without recurrence of the tumour.

Case Presentation

An 84 year old male presented to the Otolaryngology-Head and Neck Surgery service at the Queen Elizabeth II Health Sciences Centre with a left supraglottic tumour. The tumour had been present for fourteen years and had previously been asymptomatic. Recently, the patient had noticed upper airway compromise, especially during sleep. In addition, dysphagia and dysphonia were noted. On laryngoscopic examination, a large supraglottic mass was evident and the overlying mucosa appeared intact. The patient's past medical history was unremarkable except for well-controlled atrial fibrillation and minor coronary artery disease.

The plan for management was complete excision by endoscopic CO_2 laser. Informed consent was obtained preoperatively and the procedure was performed under general anaesthesia. In the operating room, the tumour was visualized with a Bouchayer laryngoscope. The mass occupied the left supraglottic area causing a bulge in the aryepiglottic folds, left false vocal cord and left laryngeal ventricle (Fig.1). The glottis appeared normal other than partial obstruction of the left true vocal cord. The lesion was removed by incising the overlying mucosa with the CO_2 laser and performing submucosal dissection with a combination of laser and cold steel instrumentation.

Postoperative pathological assessment identified the mass as an adult rhabdomyoma. Grossly, the 4.0 cm mass had a homogenous tan coloration, with a smooth surface in some areas and a papillated surface in others. On microscopy, closely-packed polygonal cells with abundant eosinophilic cytoplasm and cross-striations were noted. Crystalline-like structures were evident in the cytoplasm of some cells. Both cross-striations and crystalline-like structures were highlighted with phosphotungstic acid-haematoxylin (PTAH). Mitotic figures were not seen. Figure 2 shows the microscopic appearance of the resected tumour.

Discussion

Rhabdomyomas are rare benign tumours of striated muscle tissue that can be divided into cardiac and extracardiac types.[1] Cardiac rhabdomyomas are associated with tuberous sclerosis, whereas extracardiac varieties are not associated with any particular syndrome.[2] Extracardiac manifestations of rhabdomyomas may present in the female lower genital tract, mediastinum, stomach and thigh; however, approximately 70% of rhabdomyomas, found outside the heart, occur in the head and neck.[1] Rhabdomyomas are typically solitary lesions, although multifocal lesions have been described; however, there have been no reports of malignant transformation to rhabdomyosarcomas.[1,3,4]

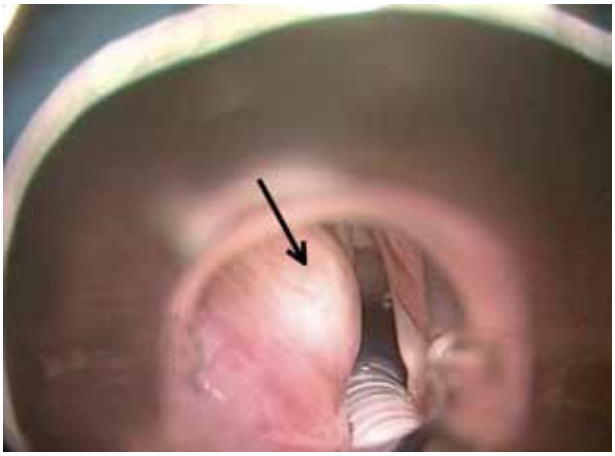

Figure 1. Intraoperative appearance of left sided supraglottic rhabdomyoma.

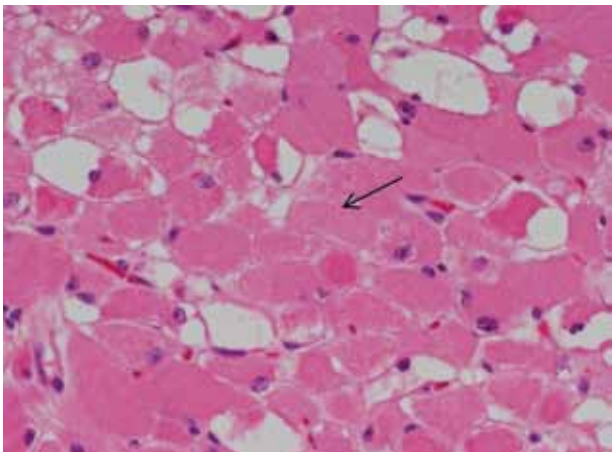

Figure 2. Photomicrograph of rhabdomyoma showing closely-packed polygonal cells with abundant eosinophilic cytoplasm. Note cell with cross striations in center of field (H&E, 200x)

Rhabdomyomas can be divided into three histological types. Adult type rhabdomyoma is found exclusively in the head and neck of adult patients. This subtype of extracardiac rhabdomyoma typically has well defined margins and is composed of closely packed, large cells with eosinophilic cytoplasm and evident cross-striations. Fetal cellular rhabdomyoma is found in the head and neck, typically in young children but may also be found in elderly men. These tumours are not well defined and may contain spindle-shaped skeletal muscle cells, few of which are mature. Fetal myxoid rhabdomyoma may be found in the lower genital tract of women and presents as a polyploidal mass with loose, edematous and myxoid stroma.[1,3] Reviews of published case reports have shown that adult type rhabdomyoma occurs most frequently.[1] In addition, extracardiac rhabdomyoma occurs more often in men than women with a mean occurrence at age 52, ranging from 16 to 82 years of age.[1,5]

Rhabdomyomas of the neck occur in close proximity to muscle tissue associated with the embryologic branchial arches.[1] This has led to the hypothesis that rhabdomyomas arise from remnants of fetal tissue that result from developmental anomalies.[1,4] However, cellular studies report cytogenetic abnormalities which suggest that rhabdomyomas are true neoplasms and not hamartomous or regenerative lesions.[6]

Symptoms of rhabdomyomas usually progress over a long duration. Commonly reported symptoms include hoarseness, foreign body sensation, dysphagia and dyspnea.[1,7,8,9] Stridor and acute airway obstruction have also been reported.[8,10]

Several cases of rhabdomyomas have been previously described, but relatively few cases have demonstrated recurrence post surgical management. Of 23 cases reviewed by Johansen et al., (1995), two cases of adult rhabdomyoma and one case of fetal rhabdomyoma did recur following surgical treatment.[4,8,11,12] Additionally, Winther et al. (1976) presented a case where tumour recurrence required reoperation on three separate occasions.[4,8] Most recently, Farboud et al. (2009) reported recurrence twice following laser excision.[13] Recurrence is thought to be due mostly to incomplete excision.[6]

From our review of the literature, there have been 32 cases of laryngeal rhabdomyoma reported. Of these, 10 cases were reported in the supraglottic space.[4,8,13-16] We present the 11th reported case of a supraglottic rhabdomyoma, and the first to be managed with laser resection without recurrence of the tumour.

In summary, adult rhabdomyoma is a rare benign tumour with insidious onset of symptoms. Symptoms range from mild to acute medical emergency. Complete surgical excision is often curative and recurrence rates are low. This, to our knowledge, is the first case of a supraglottic rhabdomyoma excised with the CO_2 laser without subsequent tumour recurrence.

References

1. Blaauwgeers JL, Troost D, Dingemans KP, et al. Multifocal rhabdomyoma of the neck. Report of a case studied by fine-needle aspiration, light and electron microscopy, histochemistry, and immunohistochemistry. Am J Surg Pathol 1989;13(9):791-9.
2. Gibas Z, Miettinen M. Recurrent parapharyngeal rhabdomyoma. Evidence of neoplastic nature of the tumour from cytogenetic study. Am J Surg Pathol 1992;16(7):721-8.

3. Helliwell TR, Sissons MC, Stoney PJ, Ashworth MT. Immunochemistry and electron microscopy of head and neck rhabdomyoma. J Clin Pathol 1988;41(10):1058-63.
4. Johansen EC, Illum P. Rhabdomyoma of the larynx: A review of the literature with a summary of previously described cases of rhabdomyoma of the larynx and a report of a new case. J Laryngol Otol 1995;109(2):147-53.
5. Kleinsasser O, Glanz H. Myogenic tumours of the larynx. Arch Otorhinolaryngol 1979;225(2):107-19.
6. LaBagnara J Jr, Hitchcock E, Spitzer T. Rhabdomyoma of the true vocal fold. J Voice 1999;13(2):289-93.
7. Liang GS, Loevner LA, Kumar P. Laryngeal rhabdomyoma involving the paraglottic space. AJR Am J Roentgenol 2000;174(5):1285-7.
8. Liess BD, Zitsch RP, Lane R, Bickel JT. Multifocal adult rhabdomyoma: A case report and literature review. Am J Otolaryngol 2005;26(3):214-7.
9. Roberts DN, Corbett MJ, Breen D, Jonathan DA, Smith CE. Rhabdomyoma of the larynx: A rare cause of stridor. J Laryngol Otol 1994;108(8):713-5.
10. Winther LK. Rhabdomyoma of the hypopharynx and larynx. Report of two cases and a review of the literature. J Laryngol Otol 1976;90(11):1041-51.
11. Hamper K, Renninghoff J, Schafer H. Rhabdomyoma of the larynx recurring after 12 years: Immunocytochemistry and differential diagnosis. Arch Otorhinolaryngol 1989;246:222-6.
12. Modlin B. Rhabdomyoma of the larynx. Laryngoscope 1982;92:580-2.
13. Farboud A, Pratap R, Helquist H, Montgomery P. An unusual cause of obstructive sleep apnoea. J Laryngol Otol 2009;123(11):e22.
14. Jensen K, Swartz K. A rare case of rhabdomyoma of the larynx causing airway obstruction. Ear Nose Throat J 2006;85(2):116-8.
15. Brys AK, Sakai O, DeRosa J, Shapshay SM. Rhabdomyoma of the larynx: Case report and clinical and pathologic review. Ear Nose Throat J 2005;84(7):437-40.
16. Pichi B, Manciocco V, Marchesi P, et al. Rhabdomyoma of the parapharyngeal space presenting with dysphagia. Dysphagia 2008;23(2):202-4.