Laryngeal spindle cell rhabdomyosarcoma in an adult

Conor M Devine, M.D.[1]; Patrick Feasel, M.D.[1,] Paul C Bryson, M.D.[1]
[1]Cleveland Clinic Foundation

Abstract

Objectives:
To describe a case of rare spindle cell rhabdomyosarcoma of the larynx.
Study Design: This was a case report and review of the literature.
Methods: An extensive review of the literature was performed searching for cases of reported spindle cell rhabdomyosarcoma in the head and neck in the adult population.
Results: The patient is a 44 year-old female diagnosed with spindle cell rhabdomyosarcoma of the larynx treated with surgical resection and chemotherapy.
Conclusions: This is the first case of laryngeal spindle cell rhabdomyosarcoma reported in the literature.

Introduction

Rhabdomyosarcoma (RMS) comprises the most common soft tissue sarcoma in the pediatric population, but is rare in adults.[1,2]
RMS is divided into three main histologic subtypes—embryonal, alveolar, and pleomorphic. Embryonal again may be divided into three main subtypes—anaplastic, botryoid, and spindle cell.[2,3]

First described by Cavazzana in 1992, spindle cell RMS is most commonly found in the paratesticular region of children. In adults, however, these are most commonly found in the head and neck.[4,5]

Histologically, spindle cell RMS is characterized by spindle cells and rhabdomyoblasts with brightly eosinophilic cytoplasm. Immunohistochemistry reveals strong desmin and myogenin positivity with a noted absence of S100 protein and keratin.[6,7]

RMS carries a relatively favorable prognosis in children, but several sources suggest a less favorable prognosis in adults. Margin status, tumor size, metastatic disease, and response to chemotherapy considered prognostic.[2,8]

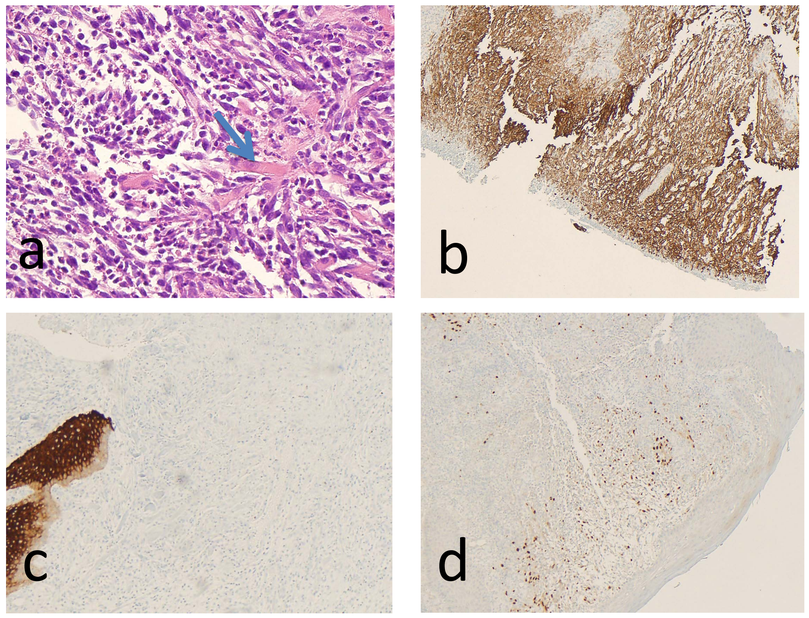

Figure 1. a) H&E stain showing prominent rhabdomyoblasts. b) Strong Desmin positivity c) Absence of cytokeratin staining in tumor stroma d) Positive myogenin staining

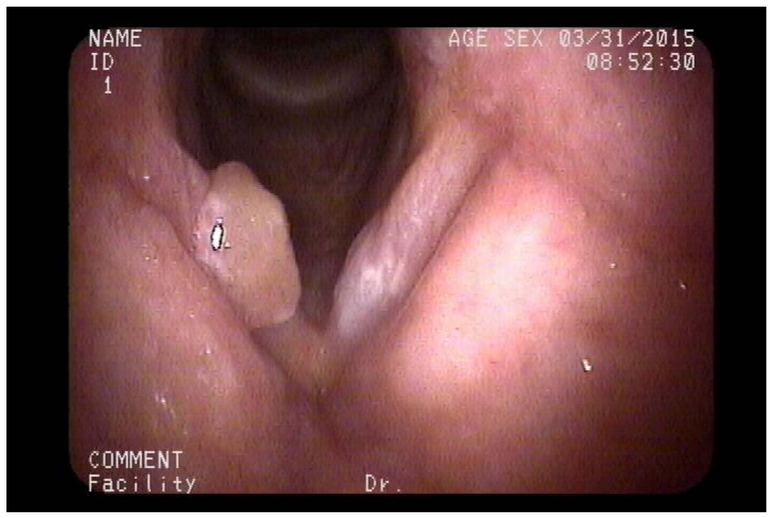

Figure 2. In-office laryngoscopy exam on intial presentation.

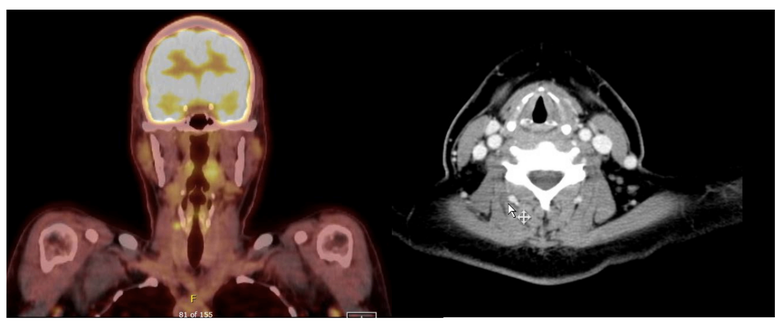

Figure 3. Immediate post-operative PET scan (left) and axial CT with contrast (right).

Case Description

A 44 year-old female presented with an approximately 3 year history of progressive hoarseness. One year prior, she was evaluated at an outside hospital where she had a right vocal fold mass biopsied. The biopsy was read as “polypoid fragment of squamous mucosa with patchy, acute inflammation and parakeratosis, submucosa with fibrosis and mild myxoid degeneration.” When her symptoms continued, she sought a second opinion and was noted to have a large smooth lesion on the mid right vocal fold that impaired closure and mucosal vibration. She was taken to the operating room for micro direct laryngoscopy and microflap excision of a 0.5cm right mid vocal fold lesion. Unexpectedly, final pathology revealed spindle cell rhabdomyosarcoma. An institutional multidisciplinary tumor board reviewed her case and recommended further excision to obtain oncologic margins as initial margins were microscopically positive. All subsequent margins were negative. She underwent CT neck with contrast and PET scan to assess for local and distant metastasis. Post operatively, she initiated treatment with 6 cycles of VAC (vincristine, dactinomycin, and cyclophosphamide). She tolerated treatment well though she developed significant mucositis from the dactinomycin. Post treatment scans have been negative for recurrance to date.

Discussion

There are several examples of laryngeal embryonal rhabdomyosarcoma in the literature, but we believe this to be the first of spindle cell variant involving the larynx. Due to the rarity of RMS, no single treatment paradigm has been established. In adults, treatment is generally aggressive multimodal therapy involving surgical resection, and chemoradiation. In this case, the primary tumor size was very small with no evidence of metastatsis and final margins were negative. Since excellent local control was obtained following surgery, a mutlidisciplinary tumor board recommended proceeding with VAC therapy alone, deferring the decision to pursue XRT until after follow up imaging had been completed. She has done well so far with no evidence of recurrent disease. Final decision concerning radiation therapy is pending.

Conclusions

Adult laryngeal rhabdomyosarcoma is a rare clinical entity with only a handful of cases in the literature. We present what we believe to be the first case of spindle cell rhabdomyosarcoma in an adult. The patient was treated with organ preservation surgical resection and chemotherapy and is currently disease free at 9 months from diagnosis.

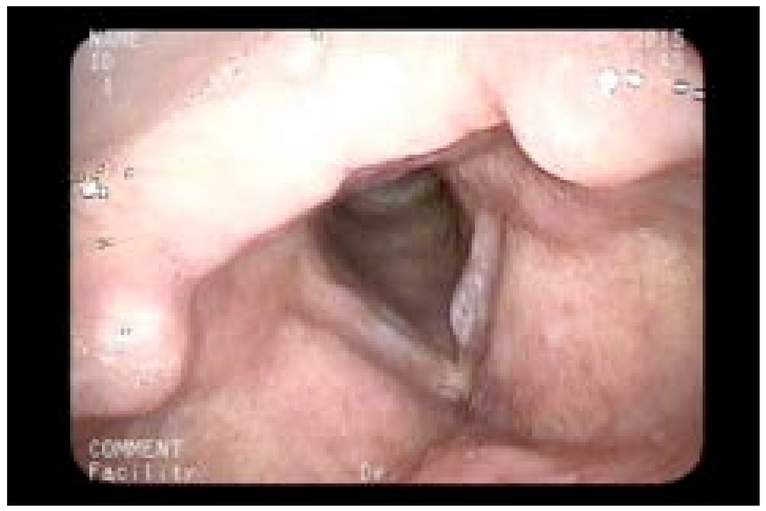

Figure 4. 1 week post operatively.

References

1. Hays D. Rhabdomyosarcoma. In Welch K, Ed. Pediatric Surgery. Chicago: Year Book Medical Publishers, Inc. 1986:276-283.
2. Hawkins WG, Hoos A, Antonescu CR, Urist MJ, Leung DH, Gold JS, Woodruff JM, Lewis JJ, Brennan MF. Clinicopathologic analysis of patients with adult rhabdomyosarcoma. *Cancer* 2001;91(4):794-803.
3. Mentzel T, Kuhnen C. Spindle cell rhabdomyosarcoma in adults: clinicopathological and immunohistochemical analysis of seven new cases. *Virchows Arch* 2006;449:554-560.
4. Cavazzana AO, Schmidt D, Ninfo V, Harms D, Tollot M, Carli M, Treuner J, Betto R, Salviati G. Spindle cell rhabdomyosarcoma. A prognostically favorable variant of rhabdomyosarcoma. *Am J Surg Pathol* 1992;16:229-235.
5. Nacimento A, Fletcher C. Spindle cell rhabdomyosarcoma in adults. *Am J Surg Pathol* 2005;29:1106-1113.
6. Rubin BP, Hasserjian RP, Singer S, Janecka I, Fletcher JA, Fletcher CD. Spindle cell rhabdomyosarcoma (so-called) in adults: report of two cases with emphasis on differential diagnosis. *Am J Surg Pathol* 1998;22:459-464.
7. Esnaola NF, Rubin BP, Baldini EH, Vasudevan N, Demetri G, Fletcher CD, Singer S. Response to chemotherapy and predictors of survival in adult rhabdomyosarcoma. *Ann Surg* 2001; 234:215-223.

Contact

Conor Devine, MD
Cleveland Clinic Foundation
Email: devinec@ccf.org
Phone: 216-444-6691