

PATIENT SPOTLIGHT

Adult Type Rhabdomyoma of the Pyriform Sinus with Cervical Esophageal Recurrence

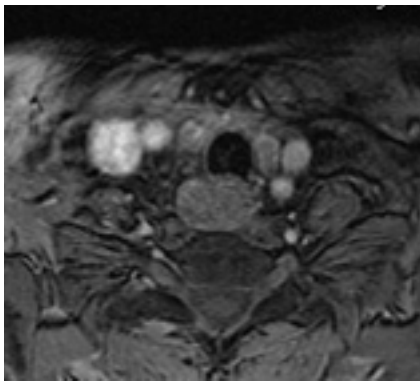


Fig 1. MRI of neck showing 2cm soft tissue mass



Fig 2. Direct laryngoscopy showing ovoid mass at the cervical esophageal opening

A 57 year-old female presented with dysphagia and frequent throat clearing.

She was found to have a right pyriform sinus mass on fiberoptic laryngoscopy that was also seen on imaging. She underwent an endoscopic laser excisional biopsy of the mass, and pathology demonstrated an adult type rhabdomyoma. After initially having symptom improvement, she again developed dysphagia, coughing, and a choking sensation. Repeat MRI revealed a 2 cm soft tissue mass in the cervical esophagus, just inferior to the cricopharyngeus (Fig 1).

Options of open resection with reconstruction for complete excision of the lesion, versus a complete or near-complete excision via an endoscopic resection were discussed. Ultimately, the decision was made to pursue an endoscopic approach in order to avoid the likely morbidity associated with an open surgical resection.

The patient was taken to the endoscopy suite in conjunction with our gastroenterology colleagues. Using a suspension rigid laryngoscope, a large smooth-walled ovoid mass was found herniating from the cervical esophageal opening (Fig 2). It was grasped and delivered further, exposing a mucosal stalk anchored at the lateral wall of the pyriform sinus. Snare cautery of the mass was used to excise it in its gross entirety (Fig 3). The tumor was a smooth, shiny, well-circumscribed, ovoid shaped, tan mass measuring 3.5 x 2.5 x 2 cm (Fig 4). Postoperatively, the patient's symptoms have improved dramatically, and she has resumed normal swallowing.



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Fig 3. Snare cautery of the mass being used for excision

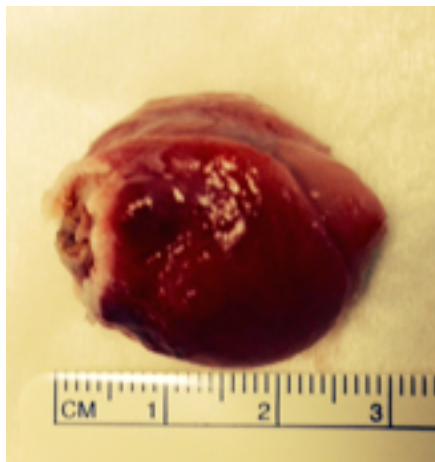


Fig 4. A smooth, well-circumscribed mass was removed measuring apex 3.5 x 2.5 x 2cm

CLINICAL REVIEW

Rhabdomyomas are benign tumors of striated muscle. They are classified by location as cardiac and extracardiac, with cardiac rhabdomyomas being more common.

The adult type rhabdomyoma (ARM) is the most common, typically presenting as a solitary mass in the head and neck region of adults. The tumor is thought to arise from the third and fourth branchial arches, which accounts for its head and neck prevalence. ARM's have been reported to occur in the oral cavity, pharynx, and larynx. As a result, symptoms are variable and can include airway obstruction, dysphagia, hoarseness, odynophagia, eustachian tube dysfunction, and aspiration.

After excision, the local recurrence rate has been estimated to be 42%, and is presumed to be due to incomplete excision. There are no reported instances of rhabdomyoma giving rise to rhabdomyosarcoma, therefore the treatment of choice remains surgical extirpation.

Though previous described cases of esophageal rhabdomyoma were treated with open surgery, they were primarily located extraluminally. In our case, the tumor was located just inferior to the cricopharyngeus muscle and was primarily endoluminal. Excision via an external approach would likely result in a permanent functional deficit in swallowing; therefore, we chose an endoscopic approach. If a future recurrence were to occur, it will likely will be amenable to another minimally invasive, endoscopic excision.



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