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WILEY
Laryngeal Reconstruction for Recurrent Desmoid Tumor Using Three-Dimensional Modeling: A Unique Approach for a Rare Tumor

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Desmoid tumors are exceedingly rare within the larynx and cause significant morbidity due to their locally aggressive and infiltrative nature. Surgery is the mainstay of treatment with previous reports describing total and near-total laryngectomy for cure. We present a case of recurrent glottic desmoid tumor managed with hemilaryngectomy and reconstructed with temporoparietal free tissue, rib, and buccal grafts. Three-dimensional modeling was utilized to optimize aerodigestive function after laryngeal reconstruction.

**Key Words:** Aggressive fibromatosis, desmoid tumor, hemilaryngectomy, laryngeal desmoid, reconstruction, three-dimensional model, 3D model.

**INTRODUCTION**

Desmoid tumors (DTs) are locally aggressive neoplasms with an impressive tendency to recur after excision. They are classified as nonmalignant due to absence of metastases or mitotic activity; however, they cause severe morbidity owing to infiltration of surrounding structures. These rare tumors occur in two to four persons per million and are exceedingly rare in the larynx, with only seven cases reported in the literature. Surgical excision with wide margins is the mainstay of treatment, with previous reports of laryngeal DTs describing total and near-total laryngectomy to obtain negative margins.

Computer-assisted design and three-dimensional (3D) modeling have been used in complex head and neck reconstruction of the maxilla, mandible, and orbit with reports of decreased operative time and improved accuracy. The complex architecture and function of the larynx offers another opportunity to utilize 3D modeling for reconstruction of hemilaryngectomy defects. We report a case using a 3D model to optimize the free tissue reconstruction of a hemilaryngectomy defect for recurrent DT of the larynx.

**CASE REPORT**

A 65-year-old male local farmer was seen with a several-month history of hoarseness and dyspnea refractory to medical therapy. He had a history of laryngeal desmoid tumor resected 15 years previously with transoral carbon dioxide (CO\textsubscript{2}) laser cordectomy and ventriculotomy.

Flexible laryngoscopy revealed a hypomobile right vocal fold with an anterior fibrotic lesion. Under general anesthesia, incisional biopsy was performed with CO\textsubscript{2} laser. While awaiting results of complex pathologic analysis, he experienced significant tumor growth and worsening of symptoms. A computed tomography (CT) scan demonstrated a poorly defined right glottic mass with paraglottic extension without destruction of adjacent thyroid cartilage, without subglottic extension, or abnormal lymphadenopathy (Fig. 1). Preoperative patient-reported outcomes for voice and swallow assessment and perceptual voice analysis were performed. These demonstrated poor voice-related quality of life and glottic function, poor perceptual assessment of voice, and confirmed normal swallow; respectively: Voice Related Quality of Life (VRQOL): 65, Glottal Function Index (GFI): 9, Grade, Roughness, Breathiness, Asthenia, Strain (GRBAS): 14, and Eating Assessment Tool 10 (EAT-10): 0.

After review of the patient’s case at multidisciplinary head and neck tumor board, decision was made to proceed with hemilaryngectomy and postoperative radiation therapy. Adjuvant radiation has been shown in some studies to reduce local recurrence of DTs. It was recommended in this case given the aggressive nature of the disease, its recurrence after 15 years despite negative margins, and the likelihood that another local recurrence in this location would ultimately lead to total laryngectomy.

Free tissue transfer reconstruction was selected to optimize aerodigestive function after anticipated adjuvant radiation. Precision in laryngeal reconstruction in
open partial laryngectomy defects is critical to balance the functions of the larynx: airway, voice, and swallow. Given its success in aiding in other areas of head and neck reconstruction, using the CT data, a 3D stereolithographic (SLA) model of the anticipated surgical defect was created to aid with reconstruction (Fig. 2).

Intraoperatively, awake tracheotomy was performed for airway control. Laryngofissure was created 3 mm onto the left thyroid ala to avoid violating the tumor and to provide adequate margin. The right glottic tumor was visualized to include the vocal process of the arytenoid to the anterior commissure and from the ventricle to infraglottis with paraglottic space invasion. The subglottis and cricoid cartilage was uninvolved (Fig. 3). Dissection removed the anterior left vocal fold and entire right false vocal fold, and released the entire thyroid ala from the cricothyroid membrane. Posterior thyroid ala behind arytenoid was divided with a drill and left in situ, and the arytenoid was disarticulated from the cricoid facet. After the hemilaryngectomy was completed, the resulting defect was a 3 × 3-cm thyroid cartilage loss with preservation of a posterior strut and circumferential cricoid preservation.

For reconstruction, rib cartilage was harvested and carved to the appropriate size, and configuration was based on the SLA model. As previously described by Gilbert et al., rib cartilage was wrapped in a temporoparietal free flap (5 × 10 cm). A buccal mucosa graft (3 × 1 cm) was harvested and then sutured on the flap at the plane of the vocal fold, which was demarcated on the 3D model (Fig. 4). The temporoparietal fascia (TPF) tissue was secured to the perichondrium of the contralateral thyroid ala. A Montgomery laryngeal stent (Boston Scientific, Shrewsbury, MA) was placed between the free tissue reconstruction and the remaining thyroid cartilage to maintain endolaryngeal configuration and was secured into place with transcutaneous sutures. On postoperative day 10, the intraluminal stent was removed endoscopically. The patient subsequently passed a modified barium swallow study and was discharged home with an oral diet and tracheostomy tube.

Histopathology revealed a 2.5 × 1.2 × 1.1-cm pink/red, well circumscribed nodule abutting the thyroid cartilage with clear margins. Light microscopy showed pleomorphic spindle cell proliferation with interlacing fascicles and admixed keloid-like collagen with increased mitotic rate compared to the previously excised lesion. Immunostains showed reactivity for smooth muscle actin, focal reactivity for calponin and CD117, and no reactivity...
for beta catenin (nuclear), desmin, CD34, anaplastic lymphoma kinase, or pan-cytokeratin.

Six weeks postoperatively, the patient was taken back to the operating room for airway evaluation, minor reshaping of the graft, and removal of mild granulation with a CO2 laser, and decannulation. He then underwent planned adjuvant radiation therapy with 60 Gy. At 1 year postoperatively he had no evidence of disease (Fig. 5). The patient-reported outcomes of swallow and voice, and perceptual evaluation of voice were reassessed. He reported good swallow function (EAT10: 0), satisfactory voice with VRQOL: 75, GFI: 3, GRBAS: 10, and no respiratory concerns.

**DISCUSSION**

The aim of this article was to report a rare case of late, recurrent laryngeal desmoid tumor, specifically describing treatment utilizing 3D modeling to optimize reconstruction after hemilaryngectomy. We also reviewed the literature of laryngeal DT.

DTs are rare, benign neoplasms of myofibroblasts that infiltrate surrounding structures causing significant morbidity. Histopathology demonstrates elongated spindle cells in the background of collagen stroma and without atypia. Differential diagnoses include nodular fasciitis, leiomyoma, solitary fibrous tumor, spindle cell squamous cell carcinoma, and fibrosarcoma. The etiology of desmoid tumors is unknown, with trauma, hormonal factors, and genetics all hypothesized to play a role in their growth and development. The biologic behavior is highly variable, with periods of stability, rapid growth, and spontaneous regression reported. Patients generally present with a slow-growing, painless mass, with additional symptoms dependent on tumor location.

The aggressive but variable behavior of this nonmalignant pathology can create a patient management dilemma. Some authors advocate close monitoring in asymptomatic patients due to the potential for spontaneous regression. Patients warrant surgical resection with wide surgical margins if functional and cosmetic outcomes resulting from surgery are acceptable. Surgery as the primary treatment results in local control rates of 75% to 80%, and many studies have shown improved local control when negative margins are achieved. Recurrent tumors are amendable to revision surgery with control equivalent to primary surgery. Radiation therapy has been shown to decrease recurrence rates in the setting of both negative and positive margins and can be used as primary treatment when surgery is unacceptably morbid. In a meta-analysis by Seinen et al., patients with recurrent desmoid tumor had significantly less local recurrences after being treated with adjuvant radiotherapy over surgery alone. This outcome may be related to the more aggressive nature of recurrent disease that encouraged the use of radiation in the patient discussed here.

This case describes laryngeal DT recurring 15 years after initial resection with rapid tumor growth after repeat biopsy and supports the described variable timeline of DTs and potential association with trauma. Despite its local aggressive tendency, DT is a nonmalignant diagnosis; therefore, maintaining the patient’s aerodigestive function after resection was paramount. Gilbert et al. demonstrated good aerodigestive outcomes with vascular reconstruction after vertical hemilaryngectomy for squamous cell carcinoma. In his series, postoperative results were comparable to literature reports of functional dysphonia and normal swallowing function. In our case, perceptual assessment of voice (GRBAS) in addition to validated, patient-reported outcome measures of vocal function and swallow (VRQOL, GFI, and EAT-10) were used to evaluate function. GRBAS is perceptual grading of voice by a trained listener, which rates five voice characteristics of grade, roughness, breathiness, asthenia, and strain; higher GRBAS scores are indicative of dysphonic voice quality. The VRQOL is a 10-question survey that assesses the effect of voice on quality of life, with higher scores representing better quality of life as it relates to voice. GFI is a four-question survey that assesses how well the glottis is functioning or closing, with higher scores suggesting worsening function. The EAT-10 is a 10-question survey that measures swallowing difficulty, with a score of 0 to 3 considered normal swallowing. The patient’s postoperative evaluation revealed persistent moderate dysphonia but improved post-treatment vocal quality, with a GRBAS improved from 14 to 10. His subjective GFI rating of 3 demonstrated improved vocal function from 9 pretreatment to within normal range, and his VRQOL improved from 63 to near normal at 75 (normal ≥80). He maintained his normal swallowing function after reconstruction with an EAT-10 score of 0.

Significant advances in computer-assisted design and creation of 3D models have facilitated many aspects of complex maxillofacial reconstruction. The use of 3D models has been shown to improve surgical precision, improve functional outcomes, and to decrease total operative time for reconstruction of complex maxillofacial defects and microvascular reconstruction of the mandible. Advantages observed in these surgeries include decreased time under general anesthesia, limited wound exposure, and more predictable functional and cosmetic results. The complex anatomy of the hemilaryngeal

**Fig. 5.** (A) Intraoperative endoscopic view of the neoglottis configuration after stent removal. (B) In-office endoscopic view of the reconstruction at 1 year postoperatively.
defect, the ultimate need for precision to balance the functions of the larynx, and the small room for error create another opportunity for the use of 3D modeling. Use of 3D models to optimize complex laryngeal reconstruction has not been previously described. In the setting of this benign disease and laryngeal preservation surgery, a primary surgical goal was optimizing our patient’s functional aerodigestive outcomes. The 3D SLA model (3D Systems, Denver, CO) was integral in creating accurate dimensions of the laryngeal reconstruction. Its use also enabled accurate positioning of the neocord to ensure a functional larynx with regards to airway and phonation (Fig. 5). Furthermore, it allowed for simultaneous shaping of the rib cartilage during ablation, which aided in decreasing operative time. Use in this case was a proof of concept. Further use in laryngeal reconstruction is needed to validate true effects on operative precision, functional outcomes, and operative time.

CONCLUSION

Laryngeal DTs have a tendency to recur and can cause severe dysphonia and airway compromise owing to locally aggressive behavior. Hemilaryngectomy can be a good treatment option in cases of unilateral glottic DT to achieve local control. Preoperative planning with 3D modeling combined with anatomic reconstruction with rib graft, buccal graft, and TPF free flap can allow good voice, respiratory, and swallowing outcomes in the setting of this nonmalignant but locally invasive disease.

BIBLIOGRAPHY