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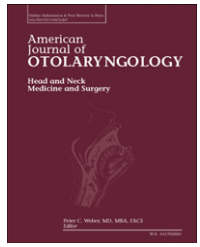

Non-epithelial tumors of the larynx: a single institution review☆

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ABSTRACT

Aim: Non-epithelial tumors of the larynx are rare and encompass a wide range of pathology. We present the decade-long experience of a single institution to define clinical presentations and outcomes.

Material and methods: This is a ten year retrospective chart review of a tertiary head and neck cancer center. Index patients were identified from a review of a pathology database, and patient demographics, presenting signs and symptoms, treatment modalities, and clinical outcomes were extracted from electronic medical records. Epithelial tumors (squamous cell carcinoma, spindle cell carcinoma, and salivary tumors), granulomas, sarcoidosis, papilloma, and amyloidosis were all excluded.

Results: Twenty-four patients with ages ranging from 2 months-old to 84 years were identified. Malignant lesions (11) included chondrosarcoma (6), Kaposi's sarcoma (2), metastatic melanoma, synovial cell sarcoma, and T cell neoplasm. Six were operated upon endolaryngeally, but four required either upfront or salvage total laryngectomy. Two received adjuvant therapy. Benign lesions (13) included hemangioma (4), granular cell tumor (3), myofibroblastic tumor (2), schwannoma (2), chondroma, and ossifying fibromyxoid tumor. Nine underwent endolaryngeal operations, and four were managed medically or with observation. None have required aggressive open resection or total laryngectomy.

Conclusion: Treatment approach of non-epithelial tumors of the larynx depends on the site and extent of the tumor, histology, and sensitivity of adjuvant therapy. Benign tumors can be managed without need for aggressive resection thereby sparing laryngeal function.

1. Introduction

Benign and malignant laryngeal neoplasms are almost exclusively epithelial in origin. The most frequently occurring malignant (squamous cell carcinoma), premalignant (squamous dysplasia), and benign (papilloma) lesions of the larynx all arise from the squamous epithelium which covers the true vocal cords. Even rare forms of laryngeal malignancies such as spindle

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cell carcinoma and salivary gland tumors are epithelial in origin. However, there is a small subset of laryngeal tumors that are non-epithelial in origin, arising from a diversity of cell types, including cartilage, muscle, or blood vessels. These tumors are usually reported as small series or isolated cases, making it difficult to assess incidence and management strategies.

In this study, we aim to present our experience with non-epithelial neoplasms over a decade at a large cancer to provide data on the incidence, management, and outcomes for these rare tumors. We further aim to provide a guide for otolaryngologists in their approach to these rare tumors by reviewing the literature to discuss approaches and treatment strategies for rarely encountered pathologies.

2. Materials and methods

In an IRB-approval protocol, the pathology archives of the Johns Hopkins Hospital and clinical records of all patients with laryngeal lesions were reviewed to identify non-epithelial laryngeal tumors between January 2004 and December 2013. Age, gender, presenting symptoms, location of the tumor, pathology, treatment modality (surgery, radiation and/or chemotherapy), and disease status were then extracted from the patient charts. Epithelial tumors (squamous cell, spindle cell, and salivary tumors), granulomas, sarcoidosis, cystic lesions, papilloma, and amyloidosis were all excluded.

3. Results

Seven hundred seventy eight (778) patient records were identified with laryngeal tumors over the decade-long study period. When all epithelial tumors (squamous cell carcinoma, spindle cell, and salivary tumors) were excluded, twenty-four patients were identified with non-epithelial tumors (Table 1).

Eleven malignant tumors were classified as non-epithelial in origin, making up 1% of all malignant laryngeal lesions. These tumors presented in adulthood, with ages at presentation ranging from 41 to 84 years old. The majority (8; 72%) were male, and the most common presenting symptom was dysphonia. Only four (36%) were smokers. The most common malignant pathology was chondrosarcoma, which presented exclusively in the cricoid cartilage. There were two cases of Kaposi's sarcoma, both in patients with long-standing HIV infections. Three lesions presented as a site of metastasis from malignancies primary to other locations – melanoma, synovial cell sarcoma, and a T cell neoplasm diagnosed in the larynx as an extra-cutaneous site from a known cutaneous malignancy.

Chondrosarcoma was the most common non-epithelial malignancy (6 patients). Initial management was divided between open and endoscopic approaches. Two patients underwent up-front total laryngectomy, two patients had endoscopic laser excisions, and one underwent an open partial resection of the cricoid cartilage. One elderly patient underwent biopsy and palliative tracheotomy and elected no further treatment. None were treated with adjuvant therapy. The recurrence rate for these lesions was high, including one patient with a total laryngectomy who then required a partial pharyngectomy and has persistent lesions in the posterior pharyngeal wall and soft palate. Both endoscopic resections had recurrence/persistence of disease, one treated with total laryngectomy and another with serial endoscopic excisions. The patient with partial resection of the cricoid ultimately required total laryngectomy. The overall rate of larynx preservation is therefore only 33%, with no patients treated with conservative surgery experiencing long-term disease-free intervals.

Both Kaposi's sarcoma patients were HIV positive, male, and smokers. A subglottic lesion caused dyspnea and was treated with endoscopic excision and adjuvant chemoradiation due to positive margins and potential for airway obstruction. Ten years of disease-free survival was achieved, with the patient ultimately expiring from issues related to the underlying immunosuppression. The lesion in the supraglottis (Figs. 1–4) caused globus sensation without airway obstruction or dysphonia, and was successfully treated endoscopically with negative margins. No adjuvant therapy was recommended.

Three patients presented with laryngeal metastases from other known primaries – melanoma, synovial cell carcinoma, and cutaneous T cell malignancy. All lesions caused symptomatic dysphonia, and were biopsied or excised endoscopically to achieve diagnosis and relieve symptoms. Malignant melanoma was diagnosed as a metastasis to the supraglottis causing airway obstruction and dysphonia in a 49 year-old woman and was completely excised endoscopically. Unfortunately the patient died within six months from the underlying disease. A lesion of the thyroid cartilage causing destruction of the laryngeal framework was diagnosed as a metastasis from mediastinal synovial cell sarcoma in a 48-year old man. Total laryngectomy was recommended but he elected treatment with combined chemoradiation and was lost to follow-up. A 60 year-old man presented with an interarytenoid mass in the setting of a cutaneous T cell malignancy. Pathology on excision was consistent with his known primary tumor and no further laryngeal treatment was required.

A total of thirteen benign non-epithelial tumors were identified in the cohort (Table 1). Five of the patients (38%) presented in childhood, with ages ranging from 2 months to 15 years-old, and age range of entire cohort was 2 months to 73 years old. The most common lesion was laryngeal hemangioma (4), followed by granular cell tumors (3), myofibroplastic tumors (2), schwannomas (2), chrondroma (1) and ossifying fibromyxoid tumor (1). Only one patient (8%) was a smoker. Eight were operated endolaryngeally with intent to treat, two were followed-up with conservative approach, and two underwent only biopsy for diagnostic purposes.

The most common benign non-epithelial tumor of the larynx in this series was hemangioma. All four presented with airway symptoms – stridor in two infants (subglottic), and exertional dyspnea in two adults (supraglottic and transglottic). The two infants were approached conservatively, one with observation where the hemangioma has been stable for five years, and the second treated with propranolol, which caused significant disease regression. Neither has required excision to maintain an adequate airway without symptoms. One of the adults has presented with upper lip and soft palate hemangiomas in addition to dyspnea, hemoptysis, and a transglottic hemangioma. He was treated endoscopically with excision for hemostasis