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Case Report

Glottic Malignant Fibrous Histiocytoma: A Case Report and Literature Review

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Keywords
Laryngeal cancer · Sarcoma · Laryngectomy · Undifferentiated pleomorphic sarcoma · Malignant fibrous histiocytoma

Abstract
Pleomorphic sarcoma of the larynx is a rare variant of laryngeal cancer. We present the case of a 59-year-old male patient who has been smoking for 40 years. He presented with signs and symptoms of an obstructive glottic mass. The diagnostic workup pointed to a malignant pathology; the histopathology report confirmed the diagnosis of glottic undifferentiated pleomorphic sarcoma (malignant fibrous histiocytoma).

Introduction

More than 170,000 patients are diagnosed with laryngeal cancer and up to 90,000 deaths occur each year; this accounts for 1–5% of all cancers and deaths annually [1, 2]. Squamous cell carcinoma accounts for 85–95% of all laryngeal cancers. It is more common in males, and multiple risk factors have been identified, tobacco and alcohol being mostly related. Most laryngeal cancers arise from the squamous epithelium, while a small percentage develops from other laryngeal tissues [1, 3, 4].

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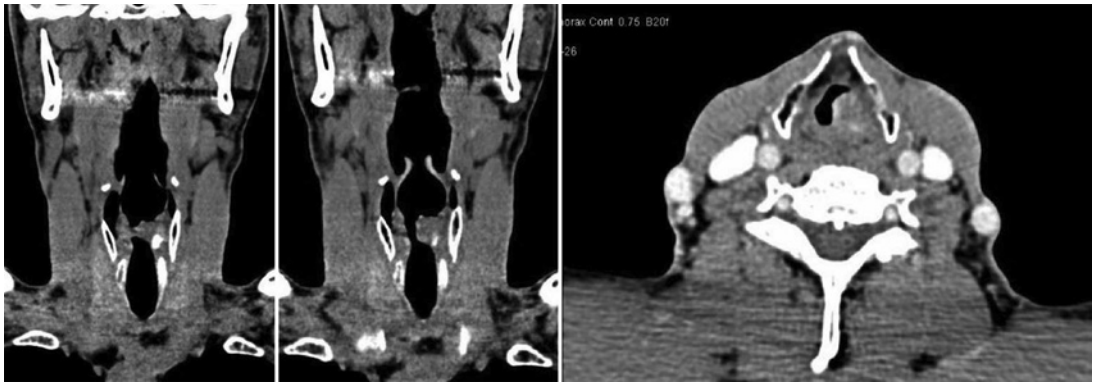

Fig. 1. Coronal and axial neck CT scan with contrast.

Radiotherapy, endolaryngeal excision, and open surgery are all accepted modalities in the treatment of early-stage glottic cancers, with endolaryngeal excision and radiotherapy having the advantage of voice function preservation. Recently, trends are shifting from open surgery toward organ preservation intervention, as there is no significant difference in survival between radiotherapy and open surgery [4].

Radiotherapy alone or in combination with chemotherapy has proven to be an effective treatment modality of laryngeal cancer in early and advanced stages. Early laryngeal cancers are treated with radiotherapy alone [5, 6], while advanced cancers require a combination of both radiotherapy and chemotherapy [7], with an adverse impact on the voice caused by glottic cancers [8].

There is only one study comparing radiotherapy to open surgery. However, interpretation of the results is limited due to concerns about the study methodology and adequacy of treatment regimens.

Case Presentation

A 59-year-old male patient, a known case of hypertension and type II diabetes mellitus on antihypertensive and oral hypoglycemic agents, with a smoking history of 40 years and a smoking index of 1,600, presented to the emergency department (ED) complaining of inspiratory stridor that started 7 days before seeking medical advice in the ED. The stridor had worsened over time until he sought medical advice. He also had a history of hoarseness of the voice for the past 5 months, weight loss of more than 10 kg (22 pounds) over the past 6 months, he has no family history of malignancies, and an unremarkable past surgical history. The fiberoptic evaluation in the ED revealed an exophytic ulcerated mass occupying the left vocal cord and partially compromising the airway with fullness of the left pyriform fossa. The right vocal cord could be seen moving. He was admitted to the surgical intensive care unit for airway observation and further evaluation.

Neck and thorax computed tomography (CT) scan with contrast (Fig. 1) showed a heterogeneously enhancing mass lesion in the left side of the larynx predominantly involving the glottic compartment and measuring approximately 1.8 × 1.5 × 1.1 cm in size. The epicenter of this mass lesion appeared to be in the left vocal cord, which extended anteromedially and caused partial compromise of the airway. There was an extension of the lesion into the left aryepiglottic fold. Superiorly, this lesion appeared to extend into the supraglottic region and reaching up to the lower part of the left pyriform sinus. Inferiorly, the mass lesion extended

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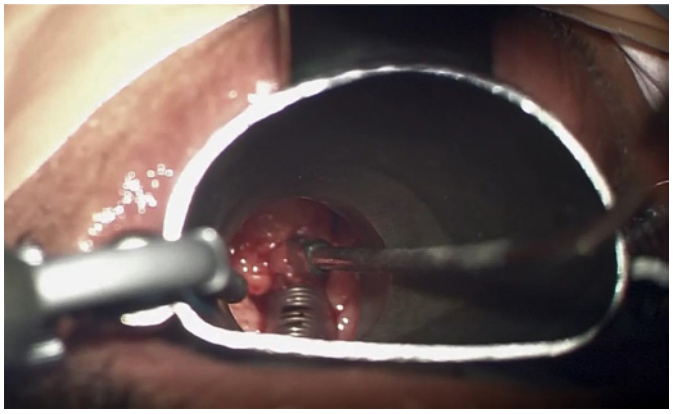

Fig. 2. Intraoperative image of the mass as it appears under micro-laryngoscopy.

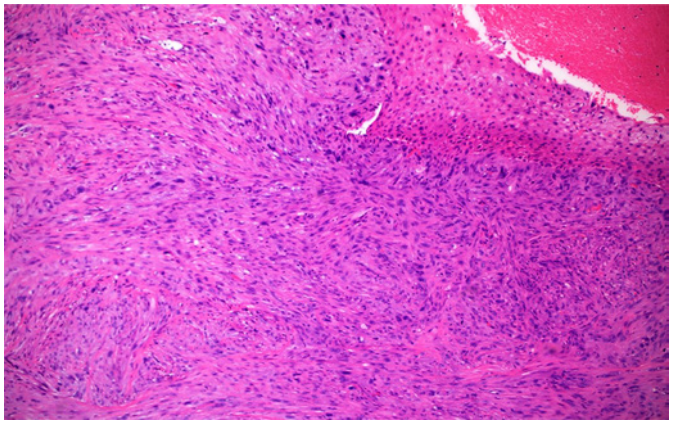

Fig. 3. Low-power view of the tumor composed of pleomorphic malignant spindle cells. Note the overlying non-atypical squamous mucosa on the top right aspect of the image (HE, ×10).

into the upper part of the infraglottic compartment. Laterally, there was extralaryngeal extension into the paralaryngeal space seen abutting the thyrohyoid membrane.

The patient underwent micro-laryngoscopy (Fig. 2) with biopsy and debulking of the mass. Intraoperatively, the tumor was fungating, arising from the supraglottic region; it involved the left vocal cord, sparing the anterior commissure, left arytenoid, left pyriform fossa, and the far posterior aspect of the left vocal fold. Specimen histopathology showed "fragments of partly ulcerated squamous mucosa with extensive underlying infiltration by a malignant neoplasm, composed of pleomorphic spindle cells, interspersed by numerous histiocytes." The malignant spindle cells exhibit frequent mitosis, including atypical mitosis; immunohistochemical stains were positive for smooth muscle actin (SMA), vimentin, and CD68 (in histiocytes), findings consistent with undifferentiated pleomorphic sarcoma (Fig. 3, 4).

Metastases workup was carried out and showed no signs of distant metastasis; the treatment plan was discussed by the multidisciplinary team, and a decision was made to do total laryngectomy with postoperative radiotherapy. The patient underwent total laryngectomy with neck dissection. Frozen sections were done; all margins were negative for malignancy. He received 6 cycles (sessions) of radiotherapy (50–60 Gy) each. On 1-year follow-up, there were no signs of local or distant disease recurrence.

On pathology, the debulked fungating tumor showed partly ulcerated squamous mucosa with an extensive underlying infiltration by a pleomorphic malignant spindle cell neoplasm. It exhibited frequent mitoses; there was no surface squamous dysplasia and no histologic evidence of the tumor originating from the surface. The tumor was subjected to a wide panel

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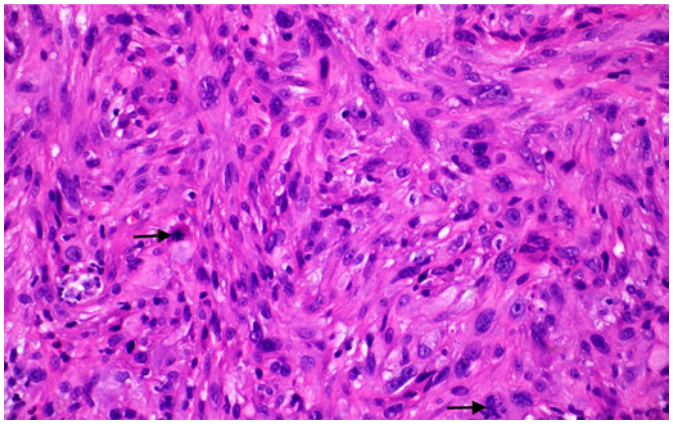

Fig. 4. High-power view of the tumor showing marked nuclear pleomorphism with frequent mitoses (black arrow) (HE, ×40).

of immune stains, but no definite cell lineage was identified. Areas of SMA and vimentin positivity were present, but various other markers, including cytokeratins, P40, P63, desmin, CD34, CD31, MyoD1, WT1, calretinin, Sox10, S100, MDM2, and CDK4, were all negative. Scattered CD68 positivity highlighted histiocytes interspersed among the tumor cells; the morphological features, coupled with the lack of specific immunohistochemical markers, were consistent with a diagnosis of undifferentiated pleomorphic sarcoma.

Examination of the subsequent laryngectomy specimen revealed residual Grade 3 (following the French Federation of Cancer Centers Sarcoma Group [FNLCC] grading system) tumor in the left vocal cord, measuring 1.5 cm in maximum dimension, with negative margins.

Discussion

Sarcomas are rare and complex soft tissue tumors of mesenchymal origin; undifferentiated pleomorphic sarcoma, known also as malignant fibrous histiocytoma (MFH), is the most prevalent type in adults; the incidence is high in the trunk and upper and lower limbs in patients between 50 and 70 years of age [9–11]. The median age of head and neck sarcoma incidence is 55–59 years [12]. Head and neck sarcomas are uncommon, with laryngeal sarcoma considered as a rare tumor, and not many cases are reported in the literature [9]. Sarcomas have different arrays of clinical presentation which varies from small slow-growing lesions to invasive ulceration and wide histologic subtypes [13].

Usually, clinical presentation depends on the site of the primary lesion and growth extent, hoarseness of the voice being the first presenting symptom in tumors originating in the larynx. Dyspnea and stridor are late symptoms, while dysphagia is unlikely unless the tumor extends into the hypopharynx [13].

TNM staging of head and neck sarcomas differs from sarcomas that arise in other parts of the body [14]. Histopathologic grade plays a significant prognostic role in soft tissue sarcomas [15]. Head and neck sarcomas carry a worse prognosis and survival rates lower than 5 years compared to extremities and superficial trunk sarcomas as shown in one single-institution series [13]. Laryngeal sarcomas carry a better prognosis than other head and neck sarcomas in general [16], but multiple factors were identified to play a role in overall survival and prognosis.

Head and neck sarcoma treatment varies; surgical excision with or without postsurgical chemoradiotherapy or chemoradiotherapy alone is chosen depending on multiple tumors and patients' related factors. Achieving a complete resection with a negative margin in head

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and neck sarcomas might be difficult due to narrow anatomic spaces and proximity to vital neurovascular structures [17]. A single study in 10 patients with laryngeal sarcomas, where 9 of them were treated with surgery and 1 patient received the full course of radiotherapy due to early diagnosis and old age at presentation, patients treated with surgery showed no evidence of disease recurrence and a 5-year disease-specific survival of 90% [17].

Management of laryngeal sarcomas will depend mainly on biologic behavior and histologic subtype of the primary tumor, while sarcomas that tend to present as a slowly growing pedunculated mass are less likely to infiltrate into deep tissue, which makes them amenable to surgery within a longer period after diagnosis, unlike squamous cell carcinoma [17]. MFH is a high-histologic-grade tumor and usually carries a poor prognosis. Moreover, high-grade tumors tend to have disseminated disease even without evidence of local recurrence, hence adjuvant chemoradiotherapy is warranted in the treatment plan [18].

Surgical intervention remains superior to any other interventional modality used in laryngeal sarcoma management as shown in a systemic review of laryngeal sarcoma [16]. In our patient, adjuvant chemoradiotherapy was given due to the aggressive nature of the histologic subtype and to minimize the chances of local or distant metastasis; surgery combined with adjuvant chemoradiotherapy will improve the survival rate in patients with laryngeal MFH [19].

Conclusion

Rare tumors represent a challenge in management due to lack of data in the literature. Unpredicted behavior of these tumors warrants a careful treatment plan considering their indolent nature and possible recurrence and metastasis. With the absence of local recurrence, the management plan should be done through retrospective analysis of outcomes in similar circumstances when available, and the plan of care must consider the best interest of the patient and should include both surgeons and oncologists; in such cases, more is the best.

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Statement of Ethics

The article describes a case report. Therefore, no additional permission from our Ethics Committee was required. Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

A.A.A.: data collection, literature search, manuscript preparation; A.R.A., H.A.H., W.R., H.A.A.: manuscript preparation; M.P.: pathology slides preparation; A.J.N.: manuscript revision and submission. All authors read and approved the final manuscript.

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