Pleomorphic undifferentiated sarcoma of the vocal fold: A case report

Adham Aljariri[1], Abdulrahman Alsaleh[1], Hasan Haider[1], Mahir Petkar[1], Hussain Al-Enazi[1], Waheed Rahman[1], and Abdulqadir Nashwan[1]

[1]Hamad Medical Corporation

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Abstract

Pleomorphic sarcoma of the larynx is an extremely rare variant of laryngeal cancer. We report a case of 59 years old male patient presented with signs and symptoms of obstructive glottic mass, all diagnostic workups pointing to malignant pathology and histopathology report confirm the diagnosis of undifferentiated pleomorphic sarcoma.

Keywords

Laryngeal cancer, Sarcoma, Laryngectomy, undifferentiated pleomorphic sarcoma

Key clinical massage

Surgeons and clinicians may face cases during their career life when there are no straight forward guidelines available for the management due to little cases reported throughout the literature.

BackgroundMore than 170,000 Patients newly diagnosed with laryngeal cancer cases and up to 90,000 death annually, that account 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 linked to laryngeal cancers, tobacco and alcohol being most related.[1,3,4]

With the majority of laryngeal cancers, a raise from squamous epithelium, small percentage develops from other laryngeal tissues.[1]

Radiotherapy, endolaryngeal excision and open surgery all are accepted modalities in the treatment of early-stage glottic cancer, with advantage for endolaryngeal excision and radiotherapy for voice function preservation, with a trend away from open surgery, and no significant difference in survival between radiotherapy and open surgery.[4]

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

There is only one study comparing radiotherapy and open surgery. However, the interpretation of its findings was limited due to concerns over the study methodology and the adequacy of treatment regimens.[4]

Case presentation A 59-year-old male patient, known to have hypertension and type II diabetes mellitus on oral hypoglycemic agents, with a smoking history of 40 years, smoking index 1600, the patient presented to emergency complaining of inspiratory stridor that started 7 days prior to ED presentation and worsened overtime till he seeks medical advice, Patient gave a history of hoarseness of voice for 5 months duration,

weight loss of more than 10 kilograms (22 pounds) over the past 6 months, no family history of malignancy, unremarkable past surgical history, Fiberoptic was done in the Emergency department (ED); it revealed an exophytic ulcerated mass occupying the left vocal cord partially and compromising the airway, a right vocal cord can be seen moving and fullness of left pyriform fossa, the patient admitted to the surgical intensive care unit (SICU) for airway observation and further evaluation.

Neck and thorax vomputed tomography (CT) scan with contrast *(Figure 1)* showed "heterogeneously enhancing mass lesion in the left side of the larynx predominantly involving glottic compartment and measuring approximately 1.8X 1.5 X 1.1cm in size. The epicenter of this mass lesion appears to be in the left vocal cord which extends anteromedially causing partial compromise of the airway. There is an extension of the lesion into the left aryepiglottic fold. Superiorly this lesion appears to extend into the supraglottic region and reaching up to the lower part of the left pyriform sinus. Inferiorly this mass lesion extends into the upper part of the infraglottic compartment. Laterally extralaryngeal extension into the paralaryngeal space seen abutting the thyrohyoid membrane"

The patient underwent microlaryngoscopy (Figure 2) with biopsy and debulking of the mass, the operative findings were fungating mass arising from supraglutic region, involves in left vocal cord, sparing the anterior commissure, left arytenoid, and left pyriform fossa, and the far posterior aspect of left vocal cord looked spared, sample sent for histopathology, pathology result was "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 SMA, Vimentin and CD68 (in histiocytes), findings consistent with Undifferentiated pleomorphic sarcoma *(Figures 3,4)* **.**

Other workups for metastasis carried out and the patient was free of distant metastasis, the patient discussed by the multidisciplinary team (MDT), plan made for total laryngectomy followed by radiotherapy, he underwent total laryngectomy with neck dissection, frozen sections done, all margins were negative for malignancy.

Histologically, the debulked fungating tumor showed partly ulcerated squamous mucosa with extensive underlying infiltration by a pleomorphic malignant spindle cell neoplasm, exhibiting frequent mitoses. There was no surface squamous dysplasia and no histological evidence of the tumor originating from the surface. The tumor was subjected to a wide panel of immunostains, but no definite cell lineage was appreciated.

Areas of smooth muscle actin (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 amongst 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 2 (following the French Federation of Cancer Centers Sarcoma Group (FNLCC) grading system) tumor in the left vocal cord, measuring 1.5cm in maximum dimension, with negative margins.

Discussion

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

Usually, the clinical presentation depends on the site of primary lesion and growth extent with hoarseness of voice being the first presenting symptom in tumor originating in the larynx, dyspnea, stridor are late

symptoms meanwhile dysphagia is unlikely unless tumor extends into the hypopharynx. [13]

TNM Staging of head and neck sarcomas differs from sarcomas arise in other parts of the body[14], also histopathologic grade plays a significant role in prognosis in soft tissue sarcomas[15], head and neck sarcomas tend to carry worse prognosis and lower 5 years survival rates than extremities and superficial trunk sarcomas as shown in one single institution series[13], laryngeal sarcoma carries a better prognosis than other head and neck sarcomas in general[16], but multiple other factors identified to play a role in overall survival and prognosis.

Soft tissue sarcoma's treatment varies based on the site, surgical excision with or without post-surgical chemotherapy/Radiotherapy, Achieving a complete resection with negative margin in head and neck sarcomas might be difficult due to narrow anatomic spaces and close proximity to vital neurovascular structures, in laryngeal cancer with extensive T3 and T4a stages total laryngectomy will achieve better survival outcomes and quality of life for the patient.[16]

Surgical intervention remains superior to any other interventional modality used in the treatment of laryngeal sarcoma as showed in a systemic review of laryngeal sarcoma.[16]

Conclusion

Rare tumors represent a challenge in management due to lack of data about them in the literature when such cases encountered; the management plan should be done through retrospective analysis of outcomes in similar circumstances, plan of care have to take into consideration the best interest of the patient and should include both Surgeons and oncologists.

Abbreviations

MDT: Multidisciplinary team

SICU: Surgical Intensive Care Unit

CT: Computed tomography

ED: Emergency Department

SMA: smooth muscle actin

FNLCC: French Federation of Cancer Centers Sarcoma Group

Declarations

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Conflict of interest

None declared

Ethics approval and consent to participate

The article describes a case report. Therefore, no additional permission from our Ethics Committee was required.

Consent for publication

The consent for publication was obtained.

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Authors' contributions

AAA: Data Collection, Literature Search, Manuscript Preparation, ARA, HAH, WR, HAA: Manuscript Preparation, MP: pathology slides preparation, AJN: Manuscript Revision and submission.

All authors read and approved the final manuscript

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Figure 1 - Coronal and Axial Neck CT with contrast

Figure 2 - Intraoperative image of the mass as appears under micro-laryngoscopy

Figure 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 (H and E x 10)

Figure 4 - High power view of the tumor showing marked nuclear pleomorphism with frequent mitoses (black arrow) (H and E x 40)

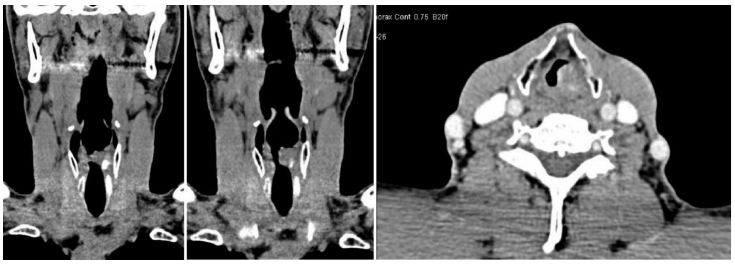


Figure 1 - Coronal and Axial Neck CT scan of the patient.

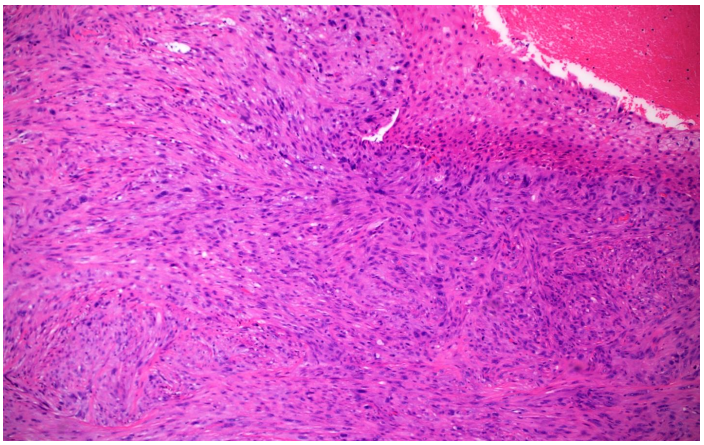
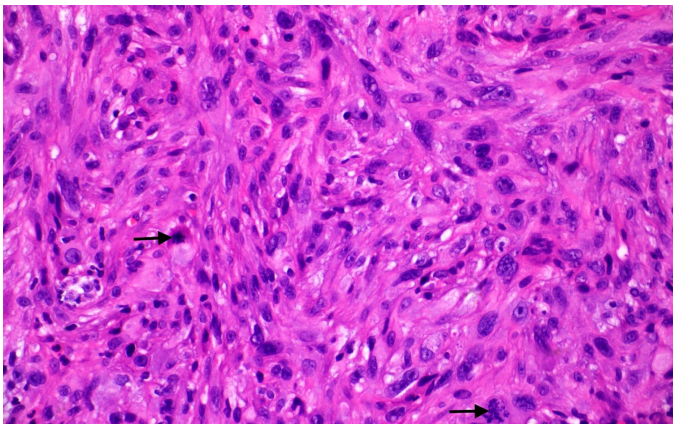


Figure 2 - Low power view of the tumor composed of pleomorphic malignant spindle cells. Note the overlying non-atypical squamous mucosa on the top right aspect of the image (H and E x 10)



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