Multi-Disciplinary Team Approach for Management of Undifferentiated Pleomorphic Sarcoma of Laryngeal Ventricle

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Received: 29 May 2023

Accepted: 3 July 2023

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DOI 10.5001/omj.2025.11

Abstract

Undifferentiated pleomorphic sarcoma of the laryngeal ventricle is extremely rare with few reported cases. We report a case of an elderly gentleman with undifferentiated pleomorphic sarcoma of laryngeal ventricle who underwent multi-disciplinary team discussion involving otorhinolaryngologist, pathologist and oncologist prior to decision of treatment. Here, we review the clinical manifestations, diagnostic criteria, and the treatment modalities for patients with laryngeal ventricle undifferentiated pleomorphic sarcoma. A multi-disciplinary team approach is extremely important for diagnosing undifferentiated pleomorphic sarcoma and providing the best treatment to achieve the best patient outcomes. This case report was written based on input from otorhinolaryngologists, pathologists, and oncologists.

Keywords: Undifferentiated pleomorphic sarcoma; Ventricles of larynx; Multi-disciplinary

Introduction

Undifferentiated pleomorphic sarcoma (UPS) of larynx is a rare malignancy with the percentage of UPS of larynx is unknown due to limited cases that have been reported.[1,2] To the best of our knowledge, there are very few studies available regarding UPS of larynx and moreover, none were reported from ventricles of larynx. Due to the paucity of cases and various behaviour of UPS tumours, it makes it difficult to analyse the best choice of treatment for UPS patients. This case report emphasizes the importance of multi-disciplinary team approach for management of UPS of laryngeal ventricle to give the best treatment option for patient.

Case Report

A man in his 70s with diabetes mellitus and hypertension presented to our otorhinolaryngology clinic complaining of hoarseness of three months duration. The patient had no difficulty in eating, and no aspiration symptoms, otalgia, shortness of breath, or neck swelling. He was not a smoker. His hoarseness

was grade 2 based on the grade, roughness, breathiness, asthenia, strain (GRBAS) scoring. Examination of the neck revealed no enlargement of cervical lymph nodes.

A 70-degree angled scope revealed a lobulated mass arising from middle third of the right vocal cord with adequate airway (Figure 1). The patient underwent endoscopic laryngeal microsurgery under general anesthesia. Intraoperatively, we observed a mass occupying almost the entire length of right ventricle. The mass was adjacent to anterior commissure but did not infiltrate it. The right vocal cord and right false cord were spared. The epiglottis, left vocal cord and false cord, arytenoids, and subglottic were normal. A decision for tumor excision and histopathological examination was made. The right ventricular mass was excised; however, we were unable to excise the tumor completely, as the base of the mass extended deep into the ventricles (Figure 2).

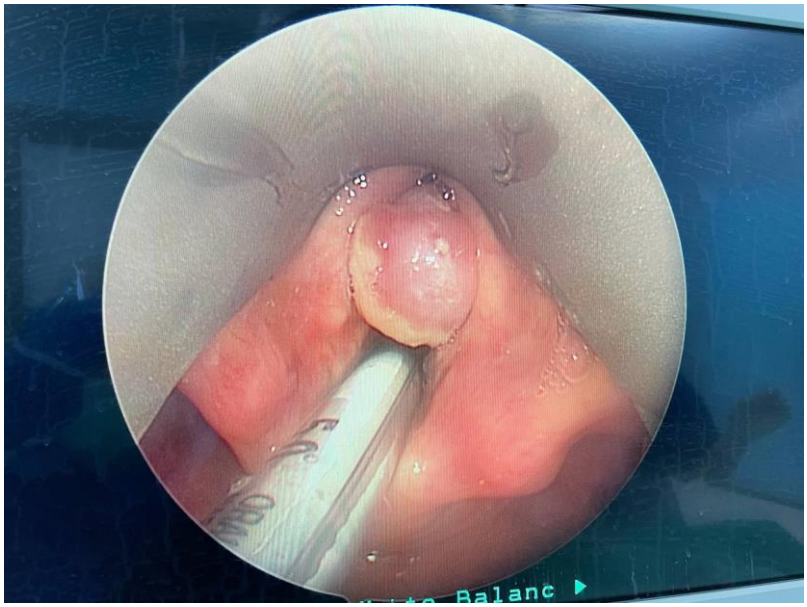

Figure 1: A 70-degree angled scope revealed a lobulated mass arising from middle third of the right vocal cord.

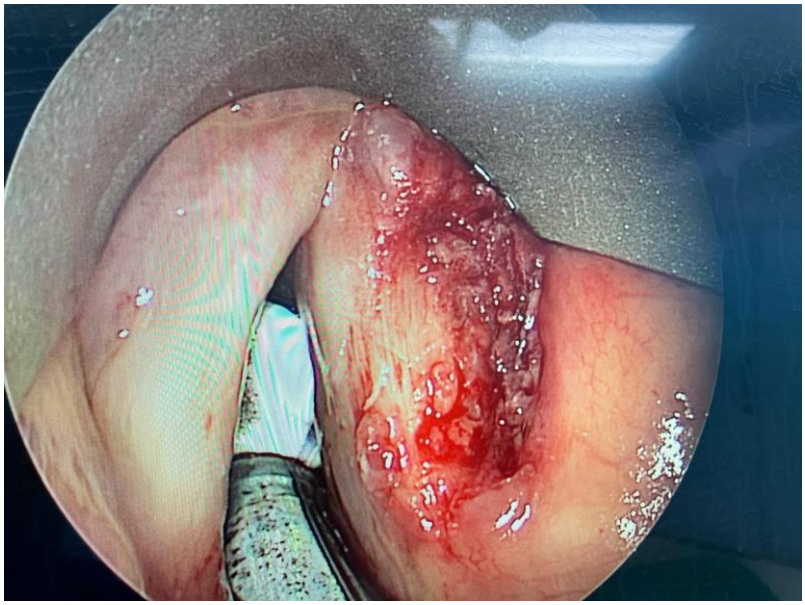

Figure 2: Intraoperatively, the base of tumor extends deep into the right ventricle.

Histopathological examination (HPE) of right laryngeal ventricular mass showed that tumor was covered with unremarkable squamous epithelium with diffuse infiltration of malignant cells within the stroma (Figure 3) and spindle cells with marked nuclear pleomorphism and abundant ill-defined cytoplasm. Multinucleation with abnormal mitotic figures were observed. The cells were strongly positive for vimentin (Figure 4) and focally positive for actin and CD34 expression. Thus, a diagnosis of UPS was reached. A deep biopsy of the ventricular mass revealed no malignant cells.

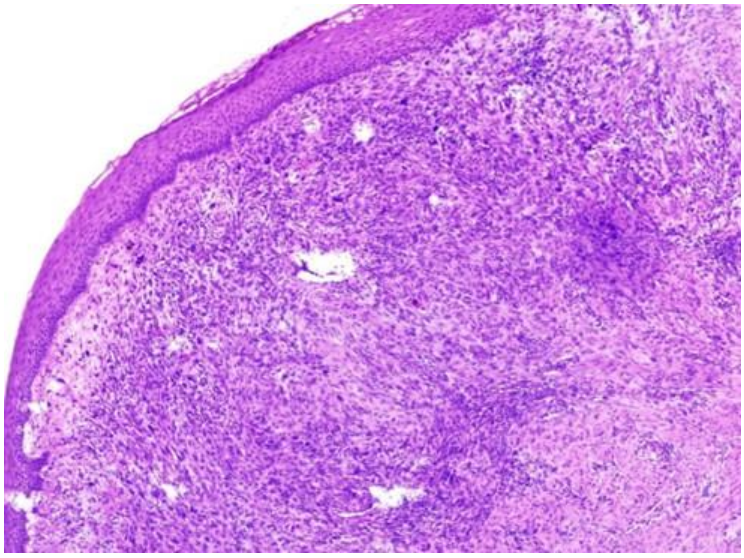

Figure 3: HPE of right laryngeal ventricular mass showed that tumor was covered with unremarkable squamous epithelium with diffuse infiltration of malignant cells within the stroma.

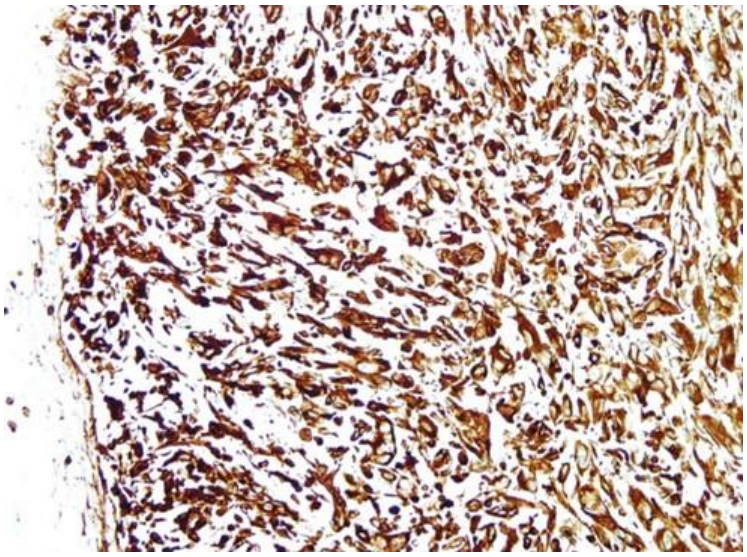

Figure 4: Immunohistochemistry test demonstrates strong positivity to vimentin.

The patient subsequently underwent a computed tomography scan from the base of the skull to the abdomen, which revealed no cervical lymph nodes or metastases.

Following discussions with the oncology team, we proceed with post-operative radiotherapy. He underwent intensity-modulated radiation therapy with a radiation dose of 66Gy in 33# over six weeks. A radiation dose of 66Gy to the tumor bed was chosen to account for possible residual disease. Owing to the supraglottic location of the tumor, bilateral cervical nodes from levels II, III, and IV were also included in

the radiation field. The ipsilateral cervical lymph nodes were irradiated with a microscopic dose of 60Gy, while the contralateral nodes received an even lower dose of 54Gy.

The patient tolerated radiotherapy treatment well, with only grade 1 mucositis and radiation dermatitis. One year after radiotherapy completion, he has minimal hoarseness. Laryngoscopic examination revealed no mass and normal laryngeal inlet structures. Patients currently undergo regular surveillance follow-up.

Discussion

UPS, previously known as malignant fibrous histiocytoma (MFH), was first reported in 1964 and represents a soft tissue sarcoma.[3,4] Due to pathological findings lacking true histiocytic origins, the term MFH was later replaced by UPS.[3] Sarcomas are uncommon malignancies of the head and neck, with laryngeal sarcomas accounting for less than one per cent of all cases.[1,2,4] UPS has variable behavior ranging from slow-growing lesions to aggressive and destructive lesions with potential for systemic metastasis.[5,6]

UPS is a diagnosis of exclusion, which makes diagnosis difficult, and at times achieved late or even missed. Thus, the role of pathologist is important to make diagnosis of UPS. A diagnosis is made when histopathological examination fails to reveal a line of differentiation after use of an adequate sample, in addition to findings from ancillary diagnostic techniques, such as immunohistochemistry.[7] Smooth muscle actin is expressed in pleomorphic myogenic sarcomas, and markers such as vimentin, p53, and Ki67 are reportedly exhibited in UPS, although they are not disease-specific.[8,9] UPS must be differentiated from other malignant tumors, such as sarcomatoid carcinoma, fibrosarcoma, myxofibrosarcoma, leiomyosarcoma, rhabdomyosarcoma, and osteosarcoma, which show a comparable degree of cellular pleomorphism.[5]

Due to the limited number of reported cases, data regarding therapeutic approaches is still limited. Multi-disciplinary approach involving otorhinolaryngologist and oncologist plays an important role in deciding on best treatment for patients with UPS. Surgery is the primary therapeutic approach for localized disease. Treatment of laryngeal UPS is based on UPS of other organs, which involves surgical excision with negative margins.[9] The therapeutic approach for laryngeal sarcomas is dictated by their size, location, and biological characteristics. Since many laryngeal sarcomas show less infiltrative characteristics, and the risk of metastasis occurs later than in laryngeal carcinomas, surgical intervention has been a primary modality of treatment.[2] Therefore, radical resection of UPS tumors is an effective modality for improving survival and decreasing recurrence. However, partial laryngectomy and laryngeal microsurgery have been described with good outcomes.[5]

Role of radiotherapy in UPS has been controversial but has evolved over the last 30 years.[2] Approximately 3–5% of UPS cases occur in patients who received radiotherapy due to other malignancies, making radiotherapy treatment controversial.[3] Sarcomas are considered radioresistant. However, Spanish Society of Medical Oncology clinical guidelines showed that perioperative radiotherapy decreases local recurrence but has no impact on survival. Post-operative adjuvant radiotherapy is considered for patients with high-grade tumors, positive surgical margins, larger tumors measuring >5 cm, and recurrent lesions.[2,10] A shared decision between otorhinolaryngologist, pathologist and oncologist must be made to give best outcome for patients.

Conclusion

We presented a rare case of UPS of the laryngeal ventricle, with the hope that it will help aid clinicians in successful diagnosis and treatment. This case presentation emphasizes the importance of each clinician including otorhinolaryngologist, pathologist and oncologist working together for the identification of UPS and for shared decision-making to provide comprehensive care for patients diagnosed with UPS.

Acknowledgments

We would like to thank Dr. Elsie Jane Anastasius helping in treating the patient.

Disclosure

The authors declare no conflict of interest.

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