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# A Case Report of Radiotherapy in Synovial Sarcoma of the Larynx: An Organ Preservation Approach

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***Authors' contributions***

*This work was carried out in collaboration among all authors. Authors SF and GM conceptualized the study and wrote and prepared the draft of the manuscript. Author AK advised on the treatment of the patient. Author OA designed the radiotherapy plan of the patient. All authors have contributed to the revision of the manuscript and have read and approved the final version of the manuscript.*

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***Case Report***

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## ABSTRACT

**Aim:** Synovial sarcoma is a malignant tumor of mesenchymal origin. It is more commonly associated with extremities in young adults. The head and neck region account for less than 10% of presentations. The larynx is an extremely rare site. We report a case of a 28-year-old male treated with laser excision followed by adjuvant radiotherapy. This study adds to the limited literature available for the management of synovial sarcoma and lay emphasis on the organ preservation treatment approach for early-stage disease.

**Case Presentation:** A 28-year-old gentleman with no comorbidities reported to us in August, 2023 with complaints of dysphagia and progressive hoarseness of voice for 2 months. He underwent

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emergency tracheostomy for stridor. Direct laryngoscopy showed a large globular mass at right aryepiglottic fold, obstructing the view of rest of the larynx. Pyriform sinus was free. The biopsy from the lesion revealed supraglottic laryngeal sarcoma. He underwent laser excision of right supraglottic lesion followed by adjuvant radiotherapy to a dose of 66 Gy in 33 fractions to post-operative bed.

**Discussion:** There are no specific guidelines for the treatment of synovial sarcoma due to its rare nature. Surgical resection with negative margins remains the mainstay of treatment, which is sometimes not achievable in the head and neck region due to the complex anatomy and the presence of major neurovascular structures. Total laryngectomy is usually preferred in these cases to provide negative surgical margins. There is severe diminution of quality of life and functional outcomes of the patient. Adjuvant radiotherapy has been used to reduce the incidence of local recurrence.

**Conclusion:** Due to paucity of literature, a multidisciplinary treatment approach is essential for the management of laryngeal synovial sarcoma and long-term follow-up is required to monitor for recurrence and improve disease-free survival. Early detection and treatment will result in laryngeal preservation thereby reducing surgical morbidity and maintaining patient's speech and swallow functions.

*Keywords: Synovial sarcoma; organ preservation; mesenchymal tumors; radiotherapy; VMAT.*

## 1. INTRODUCTION

Synovial sarcoma (SS) is rare malignancy of mesenchymal origin. It accounts for 5-10% of soft tissue sarcoma, commonly involving upper and lower limbs [1]. Approximately, around 10 % cases are reported in head and neck region, with larynx being the least common site [2].

Less than 40 cases of synovial sarcoma of the larynx are reported in the literature [3]. Similar to primary squamous cell carcinomas of the larynx, they produce symptoms due to pressure effect on larynx. This pressure effect is dependent on the size of the tumor. They might originate in any part of the larynx but tend to involve the vocal cords. Due to its rarity, treatment warrants a multi-disciplinary approach. Radical surgical excision is accepted as the treatment of choice. However, surgery also pulverises the normal anatomy of the larynx, leading to temporary or permanent physiological and functional changes, including altered airway, loss of voice, taste and smell impairments, difficulty swallowing, and appearance changes due to the tracheostoma. This in turn affects patient's mental health and quality of life [4]. We report a case of a 28-year-old gentleman diagnosed with synovial sarcoma of the larynx, treated with laser excision followed by adjuvant radiation therapy (RT) thereby preserving laryngeal function.

## 2. PRESENTATION OF CASE

A 28-year-old gentleman with no comorbidities and a chronic smoker for 8 years reported to us in August, 2023 with complaints of dysphagia and progressive hoarseness of voice for 2 months. He underwent emergency tracheostomy as he developed stridor.

Post tracheostomy routine blood investigations were sent which were within normal limits. Direct laryngoscopy showed a large globular mass at right aryepiglottic fold, obstructing view of rest of larynx. Pyriform sinus was free. The biopsy from the lesion revealed supraglottic laryngeal SS. On immunohistochemistry (IHC), the tumor cells were positive for Bcl2 and show patchy positivity for AE1/AE3, while they were negative for HMB45, desmin, MyoD1, S-100 and CD34. INI-1 showed mosaic pattern of staining in the tumor cells.

Contrast enhanced CT (CECT) scan of head and neck region showed a 3.1 X 3.7 X 4.7 cm polypoidal mass on right posterolateral aspect of supraglottic larynx, effacing right pyriform sinus. Extension of lesion to prevertebral region on right side with suspicious involvement of prevertebral fascia. No lymphadenopathy noted. PET-CT (Fig. 1) corroborated the findings of CT scan and confirmed no evidence of distant metastasis. He underwent laser excision of right supraglottic lesion. The histopathology report re-enforced the diagnosis of biphasic supraglottic synovial sarcoma, measuring 5.1 X 3.6 X 2.2 cm and all margins were free of tumor, p T3 Nx (TNM staging – AJCC 8$^{th}$ edition).

He received adjuvant RT to a dose of 66 Gy in 33 fractions to post-operative bed at 2Gy per

fraction delivered over a period of 6 weeks from September 2023 to October 2023 by Volumetric Modulated Arc Therapy (VMAT) under daily image guidance with cone beam CT scan (CBCT) in Truebeam STx (Varian Medical Systems, Palo Alto, CA). The planning target volume receiving 95% (Fig. 2a-2b) and 50% dose (Fig. 3a-3b) in the VMAT plan show good conformity in the treatment plan in axial and sagittal sections. During treatment weekly CBC and clinical review was done. He tolerated treatment well with acute toxicities of grade 2 dysphagia and grade 1 radiation dermatitis as per CTCAE version 5.

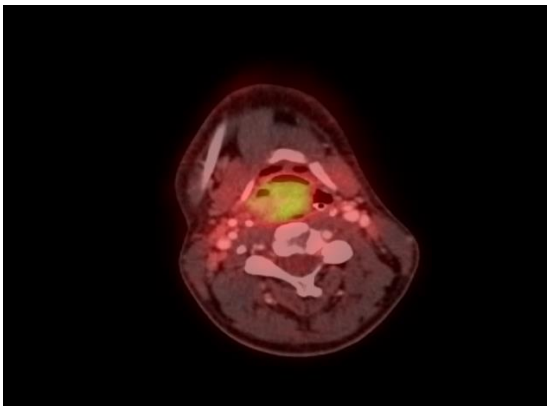

**Fig. 1. FDG avid uptake in right posterolateral aspect of supraglottic larynx with suspicious involvement of prevertebral fascia in PET-CT scan**

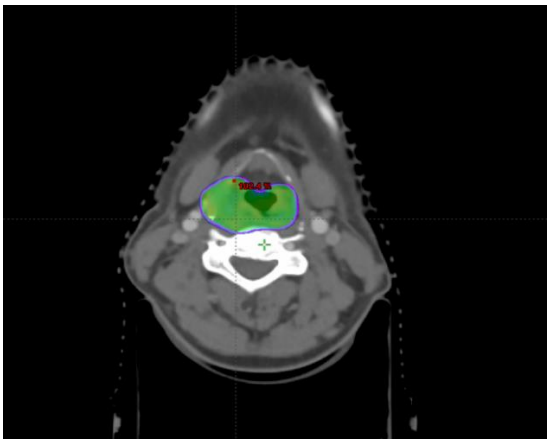

**Fig. 2a. Dose wash showing 95% coverage of planning target volume (PTV) in axial section**

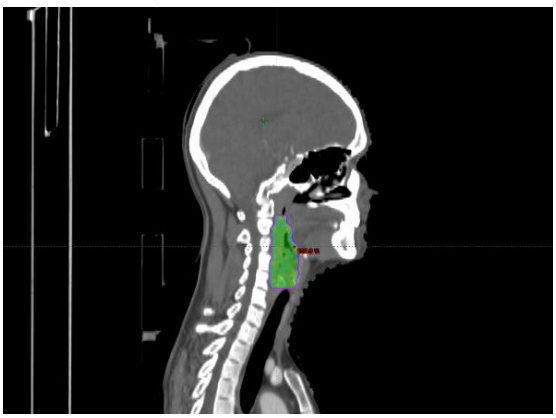

**Fig. 2b. Dose wash showing 95% coverage of planning target volume (PTV) in sagittal section**

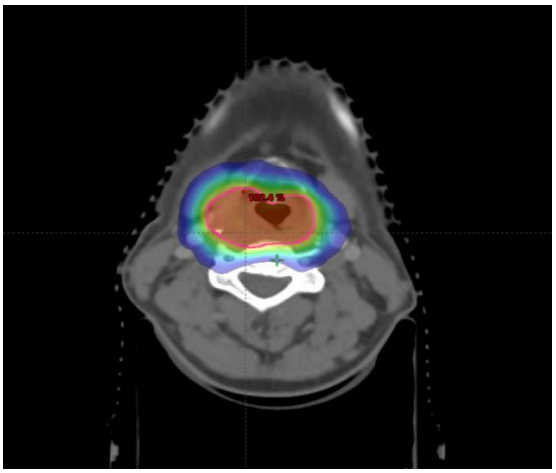

**Fig. 3a. Dose wash showing 50% coverage of planning target volume (PTV) in axial section**

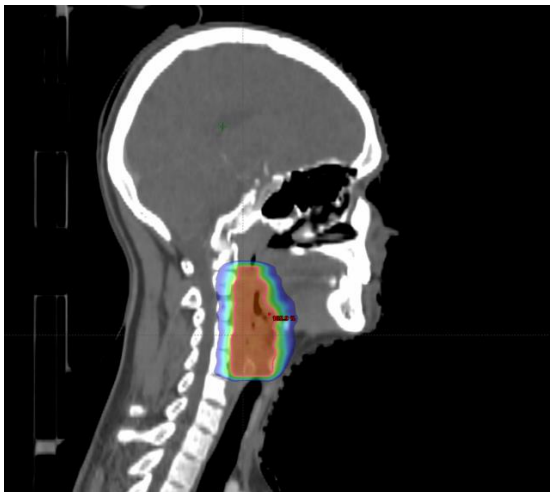

**Fig. 3b. Dose wash showing 50% coverage of planning target volume (PTV) in sagittal section**

The direct laryngoscopy done at 3 months and 8 months post treatment in January 2024 showed no residual disease. Our patient shows symptomatic improved and has been disease free till date with complete resolution of radiation side effects and does not show any late sequalae of the treatment. He has been on regular follow every 3 monthly with a laryngoscopy.

## 3. DISCUSSION

Squamous cell carcinoma is the most common malignancy of the larynx and accounts for around 90% of all laryngeal cancers [5]. Laryngeal synovial sarcoma is an extremely rare laryngeal malignancy. Histologically, it resembles a synovial membrane hence, the name synovial sarcoma [6].

The median age at diagnosis of SS is in the third decade of life, with mild male predominance [7]. A systematic review by Shein et al reported out of total 39 cases, twenty-seven (69.2%) patients were male, while 12 were female [3]. Our patient was a 28-year-old male, matching the demographic details of above cases.

Hoarseness is usually the presenting symptom followed by stridor and dyspnea. Dysphagia occurs due to the protrusion of large tumor into the hypopharynx [3]. The gross appearance is reported to be polypoidal and sometimes infiltrative. Ulceration is rare. This is in contrast with the early ulceration seen in carcinoma of the larynx. Our patient also had a similar presentation.

The three different histologic subtypes include biphasic, monophasic, and poorly differentiated sarcoma [8]. Monophasic SS includes spindle cells only. The biphasic contains glandular

structures along with epithelial elements. Poorly differentiated SS comprises uniform, densely packed, small cells and can interspersed with other histologic subtypes [9].

Immunohistochemistry plays a crucial role in histological diagnosis. SS is positive for epithelial markers including cytokeratin, epithelial membrane antigen (EMA) and vimentin and negative for CD34 and for FLI-1 [10]. Our patient was positive Bcl2 and cytokeratin (AE1/AE3).

Owing to its rare nature and paucity of literature, there are no specific treatment guidelines for this entity. As reported by Shein et al, surgery is the mainstay modality of treatment for SS of the larynx. All 39 cases underwent primary surgery, out of which 8 patients had laser excision while rest of the patients had total laryngectomy. Laryngectomized patients might develop psychological disorders such as anxiety, depression, post-traumatic stress disorder or substance abuse due to changes in physical appearance, function, and lifestyle. Patients have to adopt different strategies to cope with the threat of the disease itself as well as the immediate and long-term results of surgical treatment to return to their normal routine or achieve a new balance between their disease status and real-world life [4]. Organ preservation like laser excision of tumor helps in retaining the function and anatomy of larynx, thereby improving the quality of life of the patient. Timely diagnosis and treatment like in our case provides an opportunity for laryngeal preservation. He completed his treatment within 2 months of diagnosis.

Neck dissection (ND) is performed only in the setting of confirmed cervical lymph node involvement with no role of prophylactic lymph node dissection. However, 7 patients from the studied articles underwent an ND and none reported positive lymph nodes. Adjuvant RT has been variedly employed to reduce the incidence of local recurrence and doses in the range of 50 to 70 Gy have been reported [11].

Radiotherapy is the primary modality allowing larynx preservation in patients with tumors in this region. High-grade lesions, larger tumors (> 5 cm), positive surgical margins, and recurrent lesions are the major indications of postoperative RT [12,13]. Our patient received postoperative RT as the tumor size was more than 5 cm and due to the nature of surgery.

RT-induced laryngeal edema due to inflammation and lymphatic disruption is a common side effect. Progressive edema and associated fibrosis can lead to long-term problems with phonation and swallowing [12]. Since the primary goal of larynx preservation is speech/swallowing retention, RT-induced laryngeal dysfunction may undermine this therapeutic approach. Hence, great care should be taken in choosing the patient for organ preservation approach and providing good speech and swallowing therapy to them.

Disease recurrence is a major problem, with a local recurrence of 45% and distant metastasis in 33% of the patients with head and neck SS [14]. In locoregional recurrence treatment, repetitive radiotherapy is often not a viable option. An effective oncologic outcome can be achieved with a salvage surgery. Wound healing is impaired due to tissue fibrosis and decreased perfusion after radiotherapy. This results in a significantly increased complication rate of up to 60% with increased mortality and morbidity and significantly impaired quality of life [14,15].

In the cohort of Owosho et al. [16], there was significant effect on the disease specific survival (DSS) rates due to recurrences. The 2, 5 and 10–year DSS rates were 97%, 79% and 68%, respectively. Our patient has been disease free for 8 months without any persistent clinical radiation related toxicity.

## 4. CONCLUSION

Safety of the airway is the first concern in the advanced presentation of the SS. A multidisciplinary treatment approach is essential for the management of laryngeal synovial sarcoma. Our study emphasis the need for early diagnosis and treatment to provide a window for preservation of laryngeal function and patient's quality of life. Long-term follow-up is required to monitor for recurrence and improve disease-free survival.

## DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

## CONSENT

Written informed consent was obtained from the patient for publication of the present case report and accompanying images.

## ETHICS APPROVAL

Ethical approval not required as patient identity is not revealed.

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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