Case report

Radiotherapy in synovial sarcoma of the larynx: An organ preservation approach

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. **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 emergency tracheostomy as he developed stridor. 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 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:** Owing to its rare nature and paucity of literature, there is no specific treatment guidelines for synovial sarcoma. Surgical resection with negative margins remains the mainstay of treatment, which is not sometimes achievable in the head and neck due to the complex anatomical structure and the presence of major neurovascular structure. Adjuvant radiotherapy has been used to diminish the incidence of local recurrence. **Conclusion:** Synovial sarcoma of the larynx is a rare tumor with limited information about the treatment in the published 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.

Keywords: synovial sarcoma, organ preservation, mesenchymal tumors, radiotherapy, Volumetric modulated arc therapy

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]. Due to its rarity, treatment warrants a multi-disciplinary approach. 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).

Presentation of Case

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 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 (Figure 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 8th 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% (Figure 2a-2b) and 50% dose (Figure 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.

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.

Discussion

Squamous cell carcinoma is the most common malignancy of the larynx and accounts for around 90% of all laryngeal cancers. [4] Laryngeal synovial sarcoma is an extremely rare laryngeal malignancy. Histologically, it resembles a synovial membrane hence, the name synovial sarcoma. [5]

The three different histologic subtypes include biphasic, monophasic, and poorly differentiated sarcoma [6]. 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 [7].

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. [8] Our patient was positive Bcl2 and cytokeratin (AE1/AE3).

The median age at diagnosis of SS is in the third decade of life, with mild male predominance. [9]. 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.

Owing to its rare nature and paucity of literature, there is 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 their 39 cases had undergone primary surgery, out of which 8 patients had laser excision while rest of the patients had total laryngectomy. Neck dissection (ND) is performed only in the setting of confirmed cervical lymph node with no role for prophylactic lymph node removal. However, 7 patients from the studied articles underwent a ND and none had reported positive lymph nodes. Adjuvant RT has been variedly employed to diminish the incidence of local recurrence and doses in the range of 50 to 70 Gy have been reported. [10]

Our patient underwent a laser excision of the tumor. High-grade lesions, larger tumors (> 5 cm), positive surgical margins, and recurrent lesions are the major indications of postoperative RT. [11] Our patient received postoperative RT as the tumor size was more than 5 cm and due to the nature of surgery.

Disease recurrence is a major problem, with a local recurrence of 45% of the patient of head and neck SS and distant metastasis in 33% of the patients. [12] In the cohort of Owosho et al. [13], 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.

Conclusion

Synovial sarcoma of the larynx is a rare tumor of the head and neck region with limited information about the treatment in the published literature. Safety of the airway is the first concern in the advanced presentation of the tumor. 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.

Patient consent for publication: 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.

References

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Figures

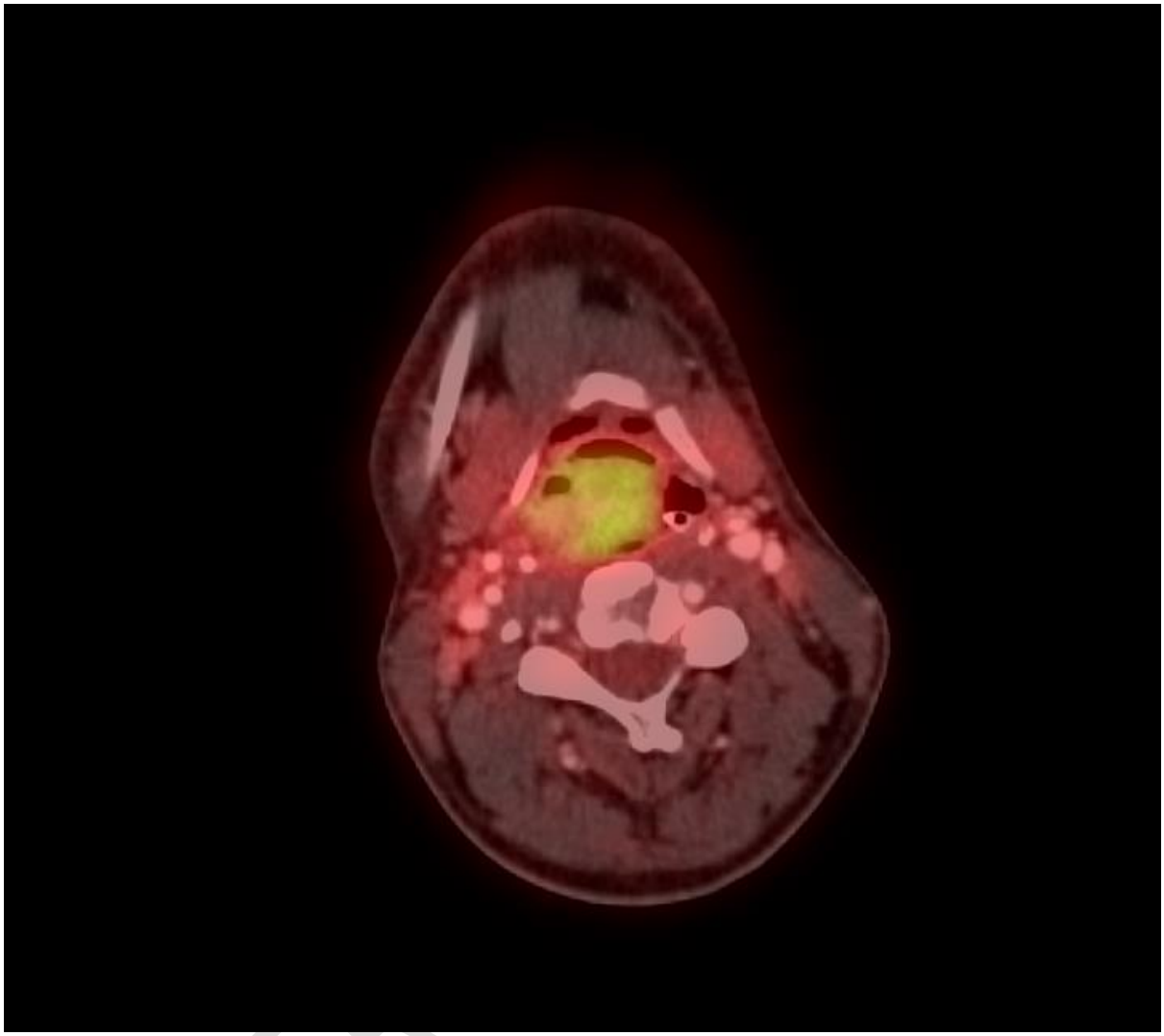

Figure 1: FDG avid uptake in right posterolateral aspect of supraglottic larynx with suspicious involvement of prevertebral fascia in PET-CT scan.

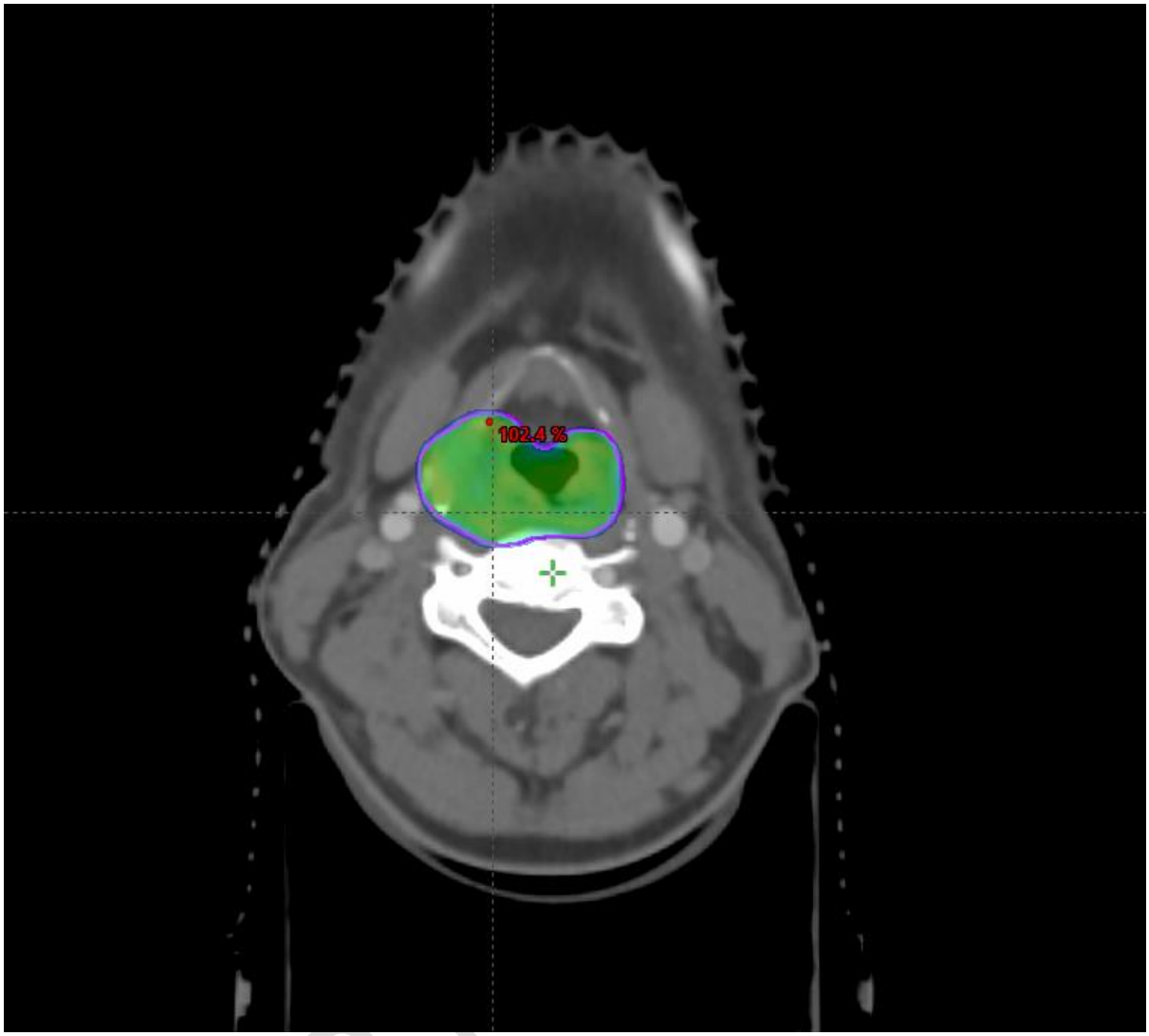

Figure 2a: Dose wash showing 95% coverage of planning target volume (PTV) in axial section.

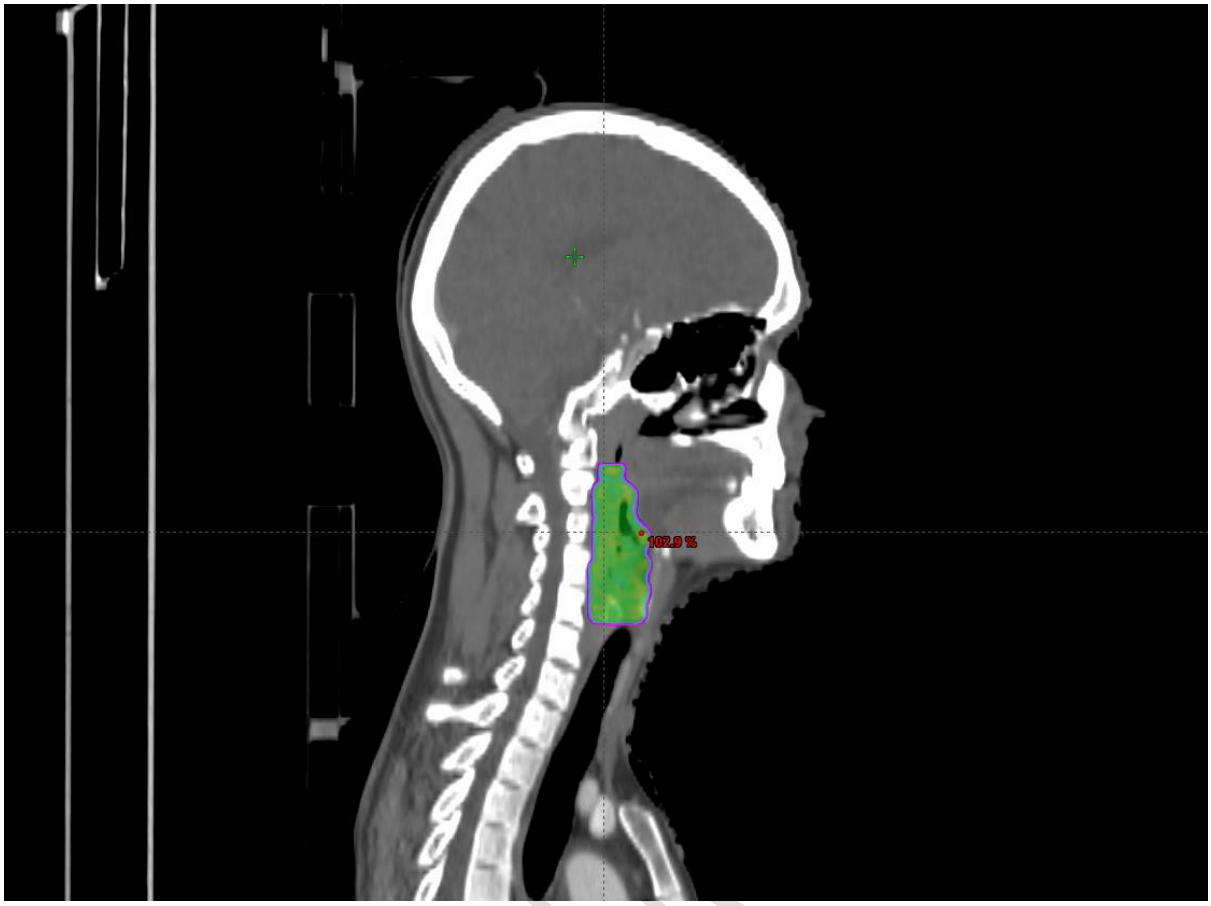

Figure 2b: Dose wash showing 95% coverage of planning target volume (PTV) in sagittal section.

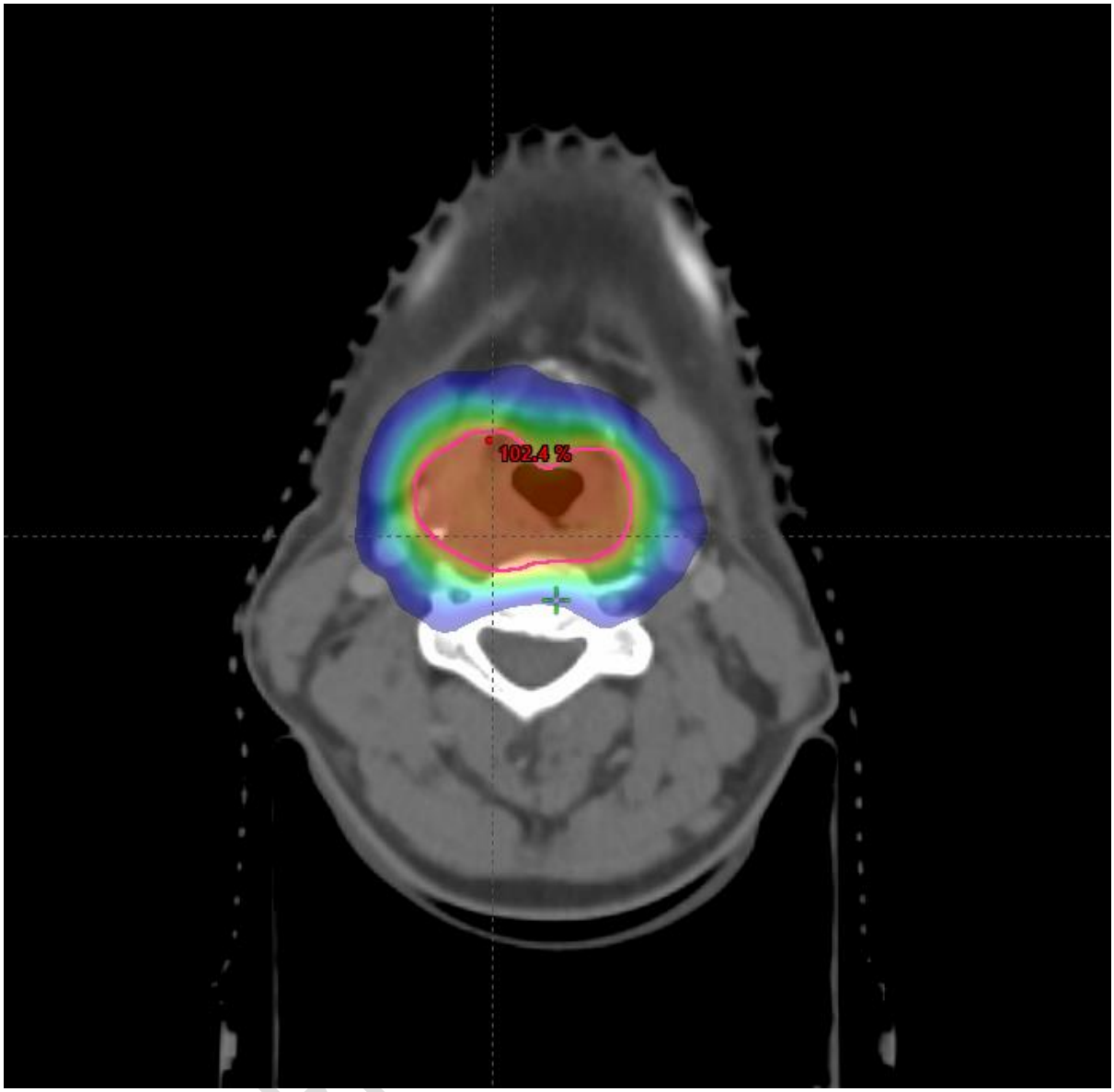

Figure 3a: Dose wash showing 50% coverage of planning target volume (PTV) in axial section.

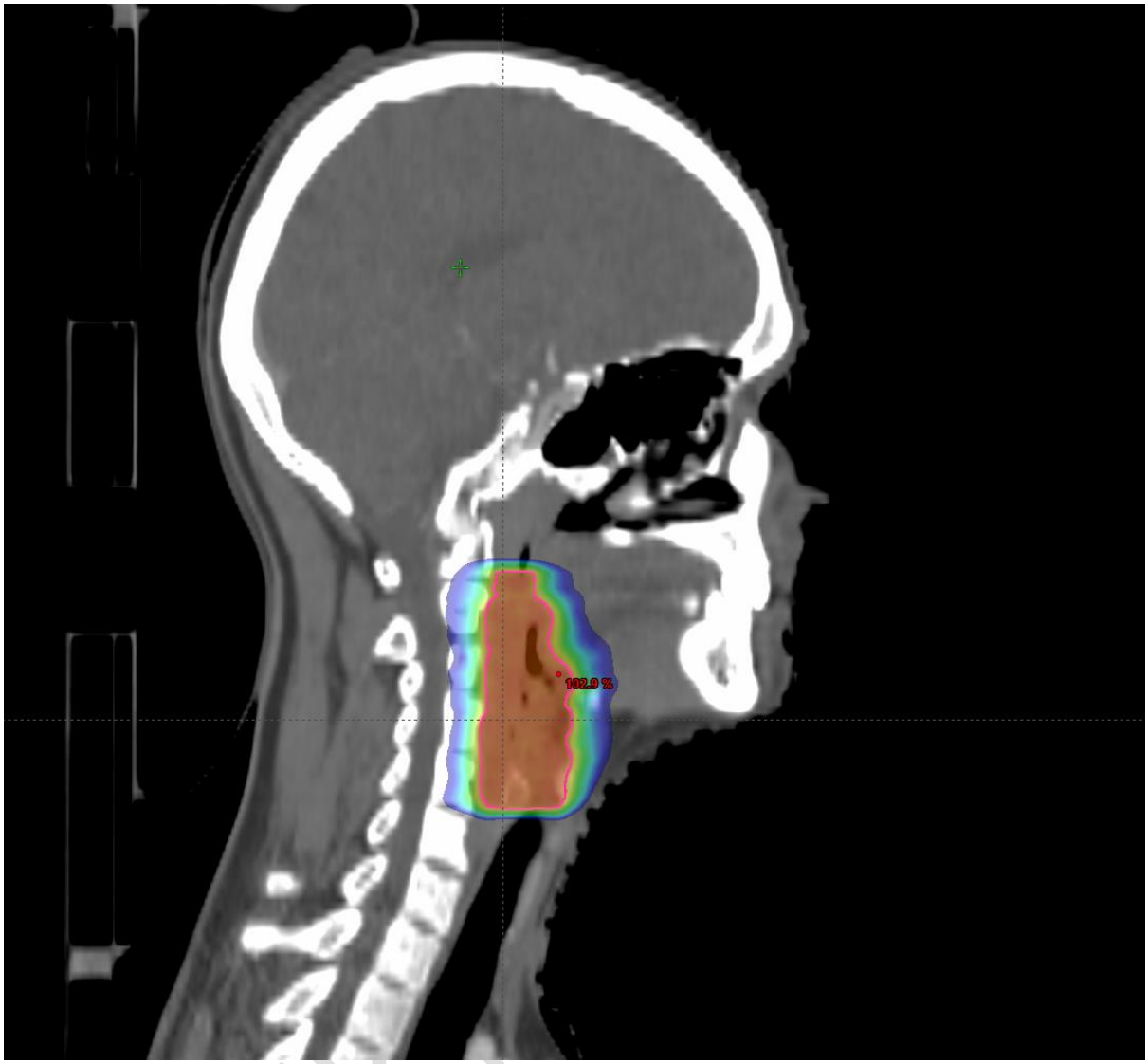

Figure 3b: Dose wash showing 50% coverage of planning target volume (PTV) in sagittal section.