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Case Report

Exceptional remission in laryngeal synovial sarcoma: a unique case treated with single-modality IMRT- Ranchi tertiary care centre experience

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ABSTRACT

Laryngeal synovial sarcoma is an extremely rare cancer originating from smooth muscle cells predominantly found in the extremities. It is uncommon in the head and neck, with laryngeal involvement particularly rare, making accurate diagnosis challenging. This case report follows the journey of a 32-year-old female patient diagnosed with laryngeal synovial sarcoma. Notably, the patient underwent an unconventional treatment using only intensity-modulated radiation therapy (IMRT). Surprisingly, the patient achieved complete remission at the one-year follow-up, making this case exceptionally rare. It is noteworthy as one of the few instances where a single treatment modality, specifically IMRT, successfully led to complete remission in laryngeal synovial sarcoma treatment.

Keywords: Laryngeal synovial sarcoma, Intensity-modulated radiation therapy, Cytokeratin, Soft tissue tumor, Spindle cell sarcoma, BCL-2

INTRODUCTION

Synovial sarcoma, an extremely rare and highly aggressive soft tissue tumor, typically originates from mesenchymal tissues. Among all the soft tissue sarcomas (STS), synovial sarcoma accounts for a relatively smaller portion, ranging from 5% to 10%. In the head and neck tumors, the occurrence of primary synovial sarcomas is exceedingly uncommon, constituting less than 5% of all synovial sarcoma cases. Among these cases, laryngeal synovial sarcomas are even more sparse. Notably, the hypopharynx is the most prevalent site for head and neck synovial sarcomas.[1] It is a translocation-associated pathognomic phenomenon characterized by t (X; 18) chromosomal translocation and eventual formation of SS18: SS X fusion oncogene, commonly affecting young adults.[2,3] The most common sites involved are the soft tissues surrounding the joints, lung, pleura, mediastinum, larynx, kidney, and buttocks.[4,5] Laryngeal synovial sarcoma demonstrates a better prognosis due to its low growth rate and a lesser probability of lymph node invasion. The malignancy presents with a male-to-female ratio of approximately 2:1, with older patients (>35 years) exhibiting worse overall survival rates. Additionally, older patient age is significantly associated with higher rates of large, invasive tumors and the presence of metastases.[3,6,7] The peculiar features include voice alterations, dyspnoea, and, in more advanced cases, stridor- a high-pitched sound caused by compromised airflow. Histologically, this malignancy exhibits a composition of spindle and epithelial cells, showcasing variations between monophasic and biphasic forms, dependent on cellular content and arrangement.[8] The conventional management for the laryngeal synovial sarcoma comprises surgical removal of the tumor, radiation, and chemotherapy.[9] The usual multidisciplinary approach to this malignancy has shown an average 5-year

survival rate in almost 70- 80% of the patients.[10] This unique case report of a 32-year-old patient with laryngeal synovial sarcoma underscores the application of a solitary treatment modality, IMRT, in managing laryngeal synovial sarcoma with careful consideration for organ preservation. Notably, the patient has achieved a remarkable 13-month remission. Attaining enduring remission in a rare and highly aggressive malignancy such as laryngeal synovial sarcoma represents a notable milestone of clinical success.

CASE REPORT

A 32-year-old female was referred to the hospital with a chief complaint of hoarseness of voice and occasional dyspnoea. Upon examination, the patient was found to be breathless with a SpO_2 level of 90% and a pulse rate of 108 beats per minute, with a gradually worsening condition leading to dyspnoea even at rest. An emergency tracheostomy was performed to alleviate the breathing distress. An abnormal mass was detected during the procedure, leading to a magnetic resonance imaging (MRI) scan. The neck MRI revealed a mass lesion measuring 2.1×1.2×1.8 cm on the left vocal cord, along with small lymph nodes bilaterally. A subsequent pathology test confirmed the diagnosis of laryngeal synovial sarcoma. Following this, the direct fibreoptic laryngeal biopsy was conducted, revealing left vocal cord palsy. The subsequent histopathological examination of the biopsy sample indicated the presence of spindle cell sarcoma, which exhibited focal positivity for cytokeratin and BCL-2 through immunohistochemical analysis (Figure 1). Standard assessments, including routine blood work, chest X-ray, and ECG (electrocardiogram), all yielded expected results. A curative approach was adopted, involving definitive external beam radiation therapy (EBRT) administered via IMRT. The patient received a total dose of 70 Gy in 35 fractions.[11]

The patient was positioned supine and immobilized with a thermoplastic mask. Axial images with 2.5 mm slice thickness were acquired from the eyebrow to the carina. After the completion of post-IMRT care, a series of awake fibreoptic laryngoscopy examinations were conducted. Acute toxicity assessment was conducted during and after the completion of treatment according to common terminology criteria for adverse events (CT CAE) v4, with the following findings: skin erythema (grade 1), pain (grade 2), dysphasia (grade 2), and laryngeal mucositis (grade 2). As a positive outcome, the patient successfully underwent decannulation three months after the radiation treatment. The patient was followed up every three months with a comprehensive clinical examination to monitor progress and detect any signs of recurrence or complications. Additionally, MRI scans were conducted every six months to assess internal healing and ensure there were no new developments or residual effects from the treatment. The patient has maintained a continuous remission for a duration of 13 months, highlighting the effectiveness of the treatment approach in both achieving remission and improving the patient's overall quality of life.

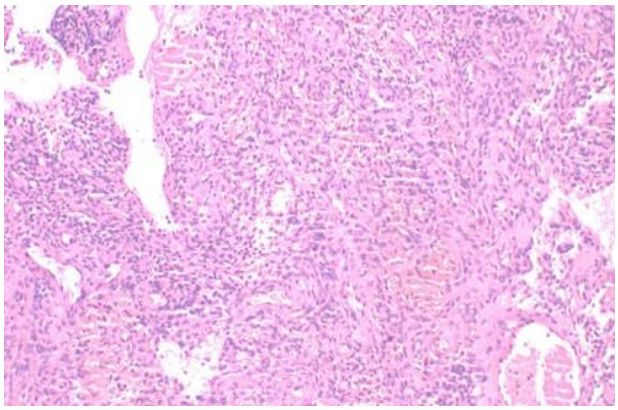

Figure 1: Spindle cells in vague fascicles with mild pleomorphism. Focal areas of necrosis were seen.

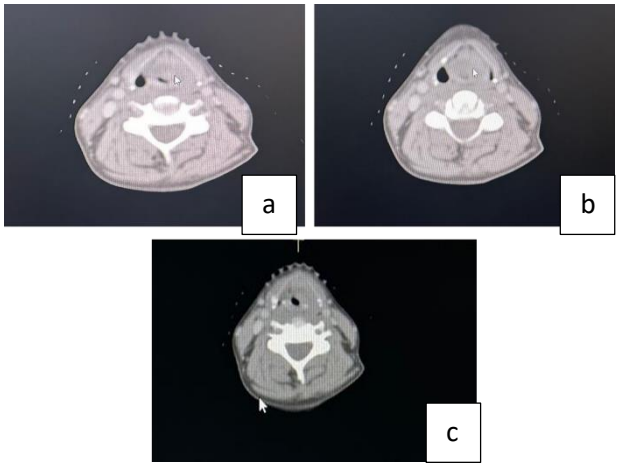

Figure 2 (a-c): MRI images of the primary tumor.

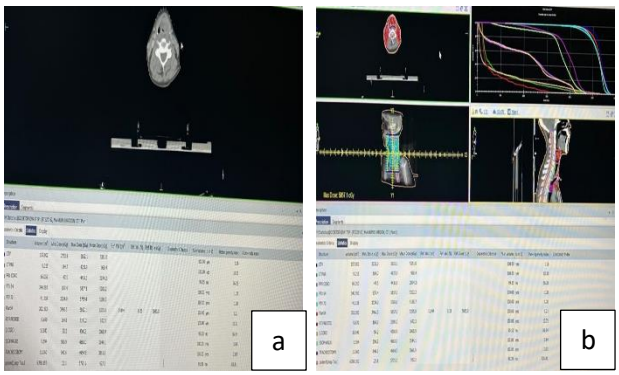

Figure 3 (a and b): Radiation therapy delineation, plan of dose distribution and dose-volume histogram.

DISCUSSION

The laryngeal synovial sarcoma occurrence is an extremely infrequent phenomenon**,** with fewer than 20 documented global cases reported in the existing medical literature. Remarkably, this malignant tumor lacks a

benign counterpart. The prevailing clinical manifestations frequently encompass dysphagia, progressive hoarseness, and exertional dyspnea as the primary complaints. Synovial sarcoma has derived its histological resemblance from developing synovial tissue; however, it is crucial to underscore that this malignancy exhibits distinctive immunophenotypic and ultrastructural characteristics that set it apart from normal synovium. Biphasic and monophasic are the two predominant histological forms of synovial sarcomas. The biphasic subtype exhibits glandular structures, prominently featuring the presence of epithelial elements, while the monophasic synovial sarcoma is characterized by the exclusive presence of spindle cells.[12] The patient in the present study presented with monophasic synovial sarcoma.

The ASTRO clinical practice guidelines (2021) suggest a multimodal treatment strategy as the optimal approach to treating synovial sarcoma.[13] Managing laryngeal sarcoma hinges on several key factors, including tumor size, location, and histological grade.[3] Typically, as suggested in the ESMO guidelines by Gronchi et al (2021), radical surgical excision stands as the cornerstone of therapy. However, due to the close proximity of the vital organs, the single modality of external beam radiation therapy (EBRT) administered via IMRT was opted in the current case as the primary treatment.[14] Notably, it is the first case reported worldwide wherein only IMRT as a single modality was administered in the laryngeal synovial sarcoma with no report of recurrence, and the patient is 13 months into remission in a study conducted by Shein et al on 39 patients with laryngeal synovial sarcoma wherein they underwent surgery to excise the tumor while 21 patients received radiotherapy as part of their initial treatment regimen in which disease recurrence was reported in 5 (12.8%) cases only.[15] Bellakhdhar et al reported two cases of laryngeal synovial sarcoma in which laryngectomy with negative margins was the treatment of choice, and it was then reported that disease recurrence posed a significant problem, with up to 45% of patients with head and neck synovial sarcoma developing local recurrence and 33% developing distant metastatic disease predominantly of the lung.[3] A comprehensive review of the existing literature on laryngeal synovial sarcoma has revealed that the survival durations, whether in the presence or absence of disease, have ranged from 3 months to 10 years. Notably, the data indicated that only 50% of patients survived beyond the 2-year mark. On the contrary, the patient in the present study, although being on a single modality, had succeeded in showing no recurrence and is in complete remission post-therapy. This outcome stands out as a remarkable and encouraging achievement, given the challenging nature of laryngeal synovial sarcoma and the varying survival rates observed in the literature.[12]

In the context of head and neck localization of synovial sarcoma, postoperative radiation therapy is commonly incorporated into the treatment regimen, although evidence regarding its efficacy in terms of local disease control or the prevention of distant metastasis remains insufficient or inconclusive.[12] However, the patient in the current study showed no distant metastasis during follow-ups.

CONCLUSION

The management of laryngeal synovial sarcoma represents a complex challenge due to the intricate anatomy and vital structures in the head and neck region. While radical surgical excision is typically the primary therapeutic approach, the case presented here illustrates a novel and successful application of a single modality, external beam radiation therapy (administered via IMRT), in the treatment of laryngeal synovial sarcoma. Notably, this is the first documented case globally where IMRT alone was utilized, and no recurrence has been observed as the patient enters the 13th month of remission.

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