

Spindle Cell Sarcoma- An Unique Entity in Larynx

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ABSTRACT

Spindle cell sarcoma of the larynx is an extremely rare malignant tumour of mesenchymal origin, with limited cases reported in the literature. Its diagnosis requires high suspicion and confirmation with histopathology and immunohistochemistry. CO₂ laser has gained popularity in laryngeal surgery due to its precision, minimal invasiveness, preservation of function, and faster recovery. The feasibility of organ-preserving CO₂ laser excision for early-stage laryngeal sarcomas is currently being explored. We report a rare case of low-grade spindle cell sarcoma of the glottis managed successfully with transoral CO₂ laser excision without the need for open surgery or adjuvant therapy, marking a valuable contribution to existing literature.

Keywords: Spindle cell sarcoma; Laryngeal neoplasm; Sarcoma of larynx; Laser assisted partial laryngectomy

INTRODUCTION

Laryngeal cancers usually arise from the surface lining of the larynx, with squamous cell carcinoma being the most common type. In head and neck cancers, spindle cell carcinomas make up about 3%, while spindle cell sarcomas of larynx are uncommon, accounting for less than 1% [1,2]. Among these rare sarcomas, chondrosarcoma is most frequently seen. Spindle cell sarcoma of the larynx presents with hoarseness, breathing difficulty, cough, and neck swelling.

The diagnosis of spindle cell sarcoma of the larynx is confirmed by clinical evaluation, imaging, and biopsy. Contrast CT or MRI scans help to determine the extent of the tumour and check for spread to surrounding tissues or lymph nodes. Histopathology shows a mix of malignant spindle cells and squamous components, which is confirmed by immunohistochemistry. Treatment involves surgical removal of the tumour, with the extent of surgery depending on the tumour size and location. Early-stage tumours may be treated with endoscopic resection or partial laryngectomy, while more advanced tumours may require a total laryngectomy followed by adjuvant therapy. We present a case of spindle cell sarcoma of the larynx with its diagnostic and therapeutic implications.

CASE REPORT

A 46-year-old male presented with progressive hoarseness of 3 months duration. Endoscopic examination revealed a tumour involving the anterior aspect of the bilateral false vocal cords, anterior half of the bilateral true vocal cords and the ventricular fold (Figure 1A and 1B). Contrast-enhanced CT neck revealed a heterogeneously hyper-enhancing well-circumscribed lesion in the anterior supraglottic region around the vestibule and extending into the ventricle (Figure 2A and 2B). The lesion involved the anterior aspect of both the false vocal cord and the right true vocal cord and is confined to the soft tissues within bilateral para glottic spaces. The patient underwent Transoral CO₂ laser-assisted type VI endoscopic cordectomy along with supraglottic extension removal (European Laryngological Society). Post operative period was uneventful and was discharged on the post-operative day 1. Histopathological evaluation was suggestive of a poorly differentiated malignant neoplasm of the larynx. Immunohistochemistry revealed that malignant cells were diffuse strong positive for SATB2, SMA and H caldesmon and negative for cytokeratin, P40, P63, Desmin, CD34, S100, myogenic, MyoD1, STAT6, SOX10, HMB45, CD21, CD68, MDM2 with a Ki67 proliferative index of 50% which was diagnostic of spindle cell sarcoma of larynx [1,4]. WBPET CT was done which revealed the absence of any distant metastasis. Multidisciplinary committee opinion was taken for the role of adjuvant therapy. However, they were of opinion that there was no role of adjuvant treatment and the patient can be observed or advised total laryngectomy. In view of age and general condition of the patient, he was advised for regular follow up and observation. Patient is on regular follow up for past 9 months without any residual or recurrence.

DISCUSSION

The most common malignancy of the head and neck region is squamous cell carcinoma (SCC), which accounts for over 90% of cases [1]. SCC also represents the vast majority of laryngeal cancers, with the glottis being the most frequently affected site [1,2]. In contrast, spindle cell sarcomas are rare tumours of mesenchymal origin and make up only a small fraction of head and neck malignancies [3]. Among the various subtypes, undifferentiated pleomorphic sarcoma (formerly known as malignant fibrous histiocytoma) which is more common in the limbs and retroperitoneum is the most frequently encountered spindle cell sarcoma [4].

The patients often present with nonspecific symptoms, the most common being progressive hoarseness due to glottic involvement [3]. Other manifestations such as dysphagia, stridor, or airway obstruction may be present depending on the tumour size and location [5]. Because of their exophytic, submucosal growth pattern, these lesions may resemble benign conditions like vocal cord polyps or granulomas, often leading to delayed diagnosis [3,4]. The endoscopic examination reveals a polypoidal or nodular lesion, most commonly affecting the true vocal cords. Narrow Band Imaging (NBI) may help delineate superficial tumour margins and assess vascularity and its application is more established in epithelial tumours [2,3].

Radiological investigations play a critical role in evaluating tumour extent and surgical planning. Contrast-enhanced CT scan is useful for assessing soft tissue involvement, airway compromise, and cartilage invasion [5], while MRI provides superior soft tissue contrast and is especially beneficial for evaluating infiltration into the pre-epiglottic and para glottic spaces [5,6]. On MRI, these tumours usually appear isointense to hypointense on T1-weighted images, and heterogeneously hyperintense on T2-weighted images, with variable post-contrast

enhancement [6]. MRI is also sensitive for detecting perineural spread, vascular encasement, and early extra laryngeal extension, which are critical for accurate staging and surgical planning.

Spindle cell sarcomas are histopathologic ally characterized by fascicles of spindle-shaped cells showing varying degrees of nuclear pleomorphism and mitotic activity [2,4]. Immunohistochemistry plays an essential role in confirming the diagnosis and excluding mimickers such as sarcomatous carcinoma [4]. Spindle cell carcinomas on the other hand demonstrate a biphasic morphology, with both epithelial squamous carcinoma in-situ or with invasion, along with subepithelial spindle cell sarcomatous lesions [1,2]. These tumours typically show negativity for epithelial markers like cytokeratin and positivity for mesenchymal markers such as vimentin, desmin, smooth muscle actin (SMA), or S100, depending on the specific sarcoma subtype [2,4].

The primary treatment is surgical resection with wide margins [2,5]. In early-stage, superficial lesions confined to the glottis, CO₂ laser excision offers a minimally invasive, organ-preserving approach with good oncologic outcomes and functional preservation [3,4]. However, larger or deeply infiltrative tumours often require open surgical procedures such as partial or total laryngectomy. Sarcomas are less radiosensitive however adjuvant radiotherapy may be indicated in high-grade tumours, close or positive margins, or when complete resection is not feasible [5,6]. Chemotherapy is usually reserved for high-grade, unresectable, or metastatic disease and is selected based on the specific histological subtype.

The prognosis of laryngeal spindle cell sarcoma depends on multiple factors including tumour grade, completeness of resection, and presence of metastasis. While early detection and complete excision can offer favourable outcomes, local recurrence and distant metastasis are not uncommon, necessitating long-term follow-up. The average 5-year survival rate is estimated to be around 50% to 70% if the tumour is detected early and removed completely with clear margins [5,6]. However, the chances of the cancer recurrence are fairly high of about 30% to 40% [5,6]. Recurrence often happens within the first few years after treatment. In view of this, close and regular follow-up is very important, especially in the early postoperative period.

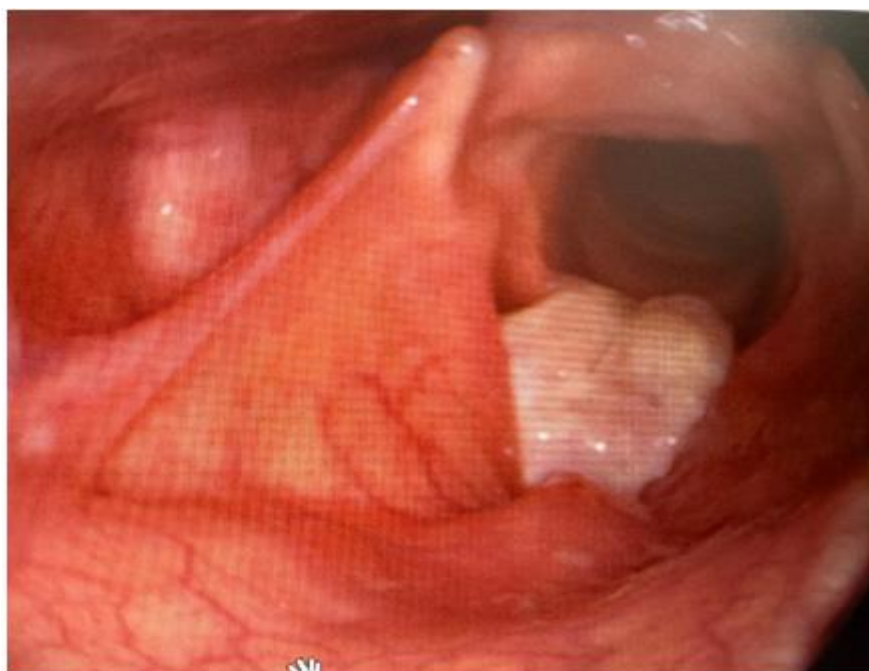


Figure 1A: Endoscopic image showing the site and extent of the lesion.

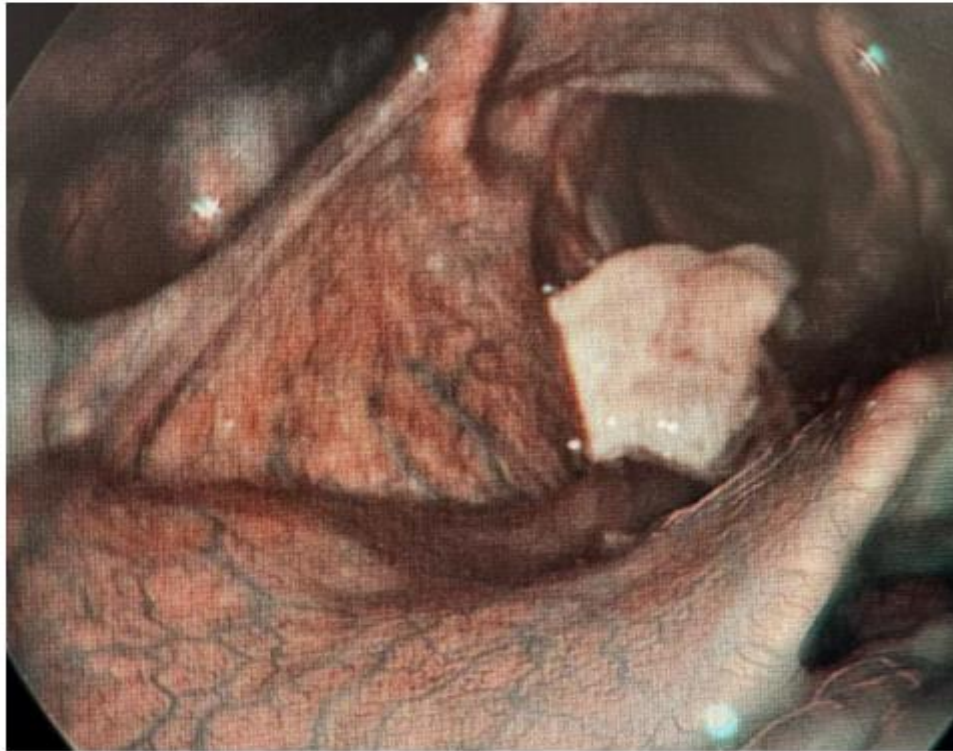


Figure 1B: Narrow band imaging of the lesion

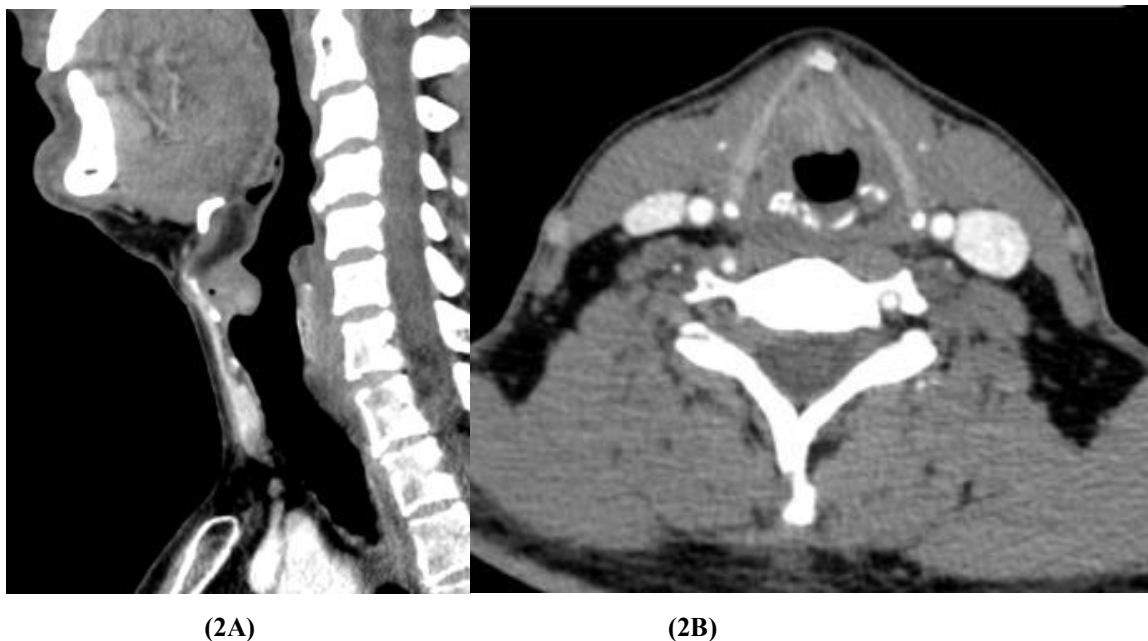


Figure 2: Contrast-enhanced CT of the neck showing a heterogeneously enhancing, well-circumscribed lesion in the anterior supraglottic region involving the supraglottic vestibule and ventricle. (A) Sagittal section. (B) Axial section.”

CONCLUSION

Spindle cell sarcoma of larynx can mimic a benign neoplasm of larynx and a detailed radiological and histopathological evaluation is mandatory for timely diagnosis and successful treatment. The rare nature of this

malignancy highlights the importance of detailed case reporting and multidisciplinary care required for its management.

DECLARATIONS

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Satish Nair: Substantial contributions to the conception or design of the work, revising it critically for important intellectual content, analysis and interpretation of data for the work and drafting the work and final approval of the version to be published.

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