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Case Report

Ear, Nose and Throat Section

Spindle Cell Carcinoma of Larynx: A Rare Case Report

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ABSTRACT

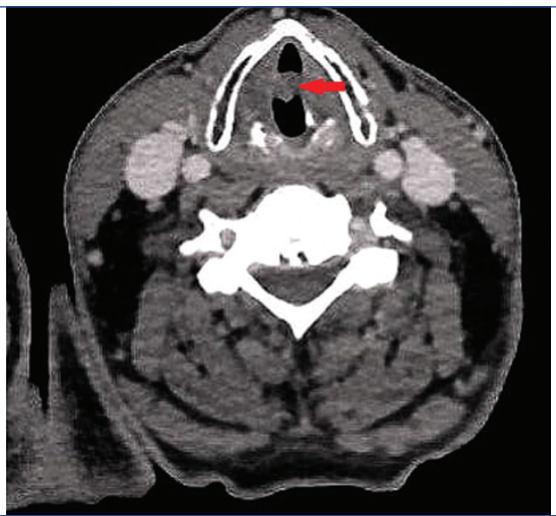
Squamous Cell Carcinoma (SCC) is the most common malignancy affecting the larynx in the Indian population. Spindle Cell Carcinoma (SpCC), representing 2-3% of all laryngeal tumours and 1% of head and neck cancers, is a rare variant of SCC that includes a mesenchymal-like malignant spindle cell component. This tumour predominantly occurs in males over the age of 65 years. Known risk factors include smoking and excessive alcohol consumption. Patients often present with hoarseness, as the glottis is the most frequently affected site in about 70% of cases. The tumour typically appears polypoid. Histopathological examination reveals that laryngeal SpCC is a biphasic tumour, consisting of both SCC and a malignant spindle cell component with mesenchymal characteristics but of monoclonal epithelial origin. The primary treatment for this condition is surgical excision, while radiotherapy is indicated for recurrent or more extensive tumours. In the present case report, the authors discussed a case of 45-year-old male patient who presented with a history of hoarseness of voice for one month. Direct laryngoscopy revealed a growth located on the anterior two-thirds of the right vocal cord, along with reduced mobility of that vocal cord. A biopsy was taken, and histopathology and immunohistochemical analysis reported the diagnosis as spindle cell variant SCC. The diagnosis is confirmed through histopathology and immunohistochemical analysis, where tumour cells express both epithelial and mesenchymal markers. The prognosis for laryngeal SpCC is generally poorer than that for laryngeal SCC, necessitating regular follow-up for affected patients. Benign laryngeal lesions that appear harmless should undergo a thorough evaluation and histopathological examination. This is crucial to identify rare conditions, such as sarcomatoid carcinoma, which can closely resemble benign lesions and require appropriate management.

Keywords: Biphasic tumour, Laryngeal spindle cell carcinoma, Neoadjuvant therapy

CASE REPORT

A 45-year-old male patient was referred to a tertiary care facility with a complaint of hoarseness that had progressively worsened over the past month. The patient had a significant history of cigarette smoking spanning 20 years. Upon examination of the head and neck region, no cervical lymphadenopathy was detected. Direct laryngoscopy revealed a growth located on the anterior two-thirds of the right vocal cord, along with reduced mobility of that vocal cord. Provisional diagnosis was carcinoma of larynx. Differential diagnosis was vocal nodules, vocal polyp, vocal cyst, laryngeal papilloma and malignant lesions.

A contrast-enhanced Computed Tomography (CT) scan of the neck identified a well-defined soft tissue lesion measuring approximately 8×5 mm. This lesion originated from the medial surface of the anterior portion of the right vocal cord and extended into the glottic airway, causing narrowing [Table/Fig-1].

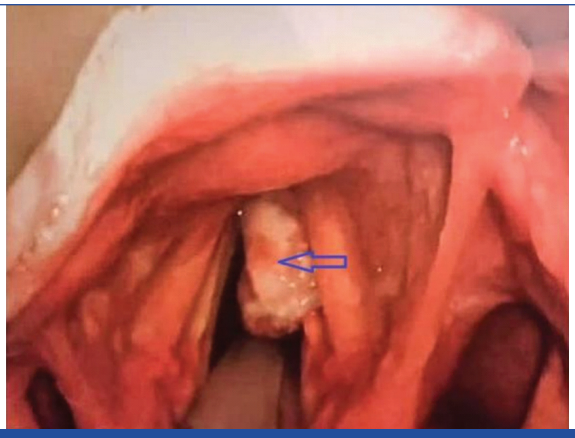


[Table/Fig-1]: CT scan image showing a well-defined, homogeneously enhancing soft-tissue density lesion measuring approximately 8x5 mm arising from the medial surface of the anterior portion of the right vocal cord, protruding into and narrowing the adjoining glottic airway.

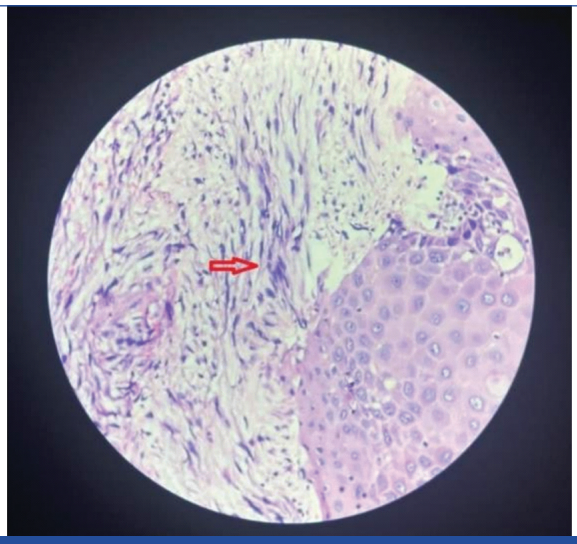
The patient underwent an examination under anaesthesia followed by wide local excision. General anaesthesia was administered, and a microlaryngeal scope was introduced and secured with Riecker's chest piece for optimal visualisation of both vocal cords and the growth on the anterior two-thirds of the right vocal cord. A polypoidal mass [Table/Fig-2] was excised using Kleinsasser cup forceps and laryngeal straight scissors. The mass exhibited a smooth surface and a soft-to-firm consistency before being sent for pathological analysis. Haemostasis was successfully achieved during the procedure.

The tumour exhibited interlacing fascicles and bundles of spindle-shaped cells characterised by hyperchromatic and pleomorphic nuclei, some of which displayed prominent nucleoli and a varying amount of cytoplasm, indicating a high-grade malignancy. Notable mitotic activity was also observed. Pathological analysis confirmed the presence of a spindle cell variant of SCC [Table/Fig-3]. The sample was subsequently subjected to immunohistochemical staining, which revealed positivity for p40 and vimentin, confirming the diagnosis of SpCC. In contrast, markers such as S100, Signal Transducer and Activator of Transcription 6 (STAT6), Erythroblast Transformation-specific-related Gene (ERG), Cytokeratin (CK) and Cluster Differentiation 31 (CD31) returned negative results. A biopsy confirmed the diagnosis as spindle cell variant SCC.

The patient's SpCC was classified as T1, as the tumour remained localised to the anterior commissure while extending into the glottic region of the vocal cord. The patient underwent microlaryngoscopy for the excision of the malignant mass from the anterior two-third of the right vocal cord. The patient received neoadjuvant therapy



[Table/Fig-2]: Preoperative picture showing a polypoidal mass on the anterior two-thirds of the right vocal cord.



[Table/Fig-3]: Histopathology specimen showing a spindle cell variant of SCC, showing a tumour consisting of interlacing fascicles and bundles of spindle-shaped cells having hyperchromatic pleomorphic nuclei, some showing prominent nucleoli and a scant to moderate amount of cytoplasm, suggesting a high-grade malignant tumour from the anterior two-third of the right vocal cord (H&E, 40x).

with cisplatin as advised by the medical oncologist after diagnosis was confirmed by the histopathology report. Following treatment, the patient experienced significant improvement in symptoms and regained effective voice control.

DISCUSSION

The majority of laryngeal malignancies, over 95%, are SCC. In contrast, Spindle Cell Carcinoma (SpCC) is a rare variant that accounts for about 1.3% of these tumours [1]. It was first reported in the literature in 1933 by Figi as 'larynx sarcoma' [2]. Due to its aggressive nature and distinct histological features, it is often referred to as a collision tumour [3]. This type of carcinoma is primarily observed in men aged 60-70 years. SpCC is closely linked to a history of cigarette smoking, and its components typically present in a nested arrangement, with the spindle cell component comprising the majority of the tumour [4].

Laryngeal SpCC typically presents with a polypoid appearance, which can lead to misdiagnosis as a laryngeal polyp when it involves the true vocal cords [5]. Imaging findings for laryngeal SpCC lack specificity, making thorough histopathological and immunohistochemical evaluations essential for accurate diagnosis. As a variant of SCC, SpCC contains a malignant spindle cell component resembling mesenchymal tissue [6]. Advances in pathological techniques have confirmed that laryngeal SpCC is a monoclonal epithelial neoplasm, with spindle cells arising from the SCC component through epithelial-mesenchymal transition [7].

The management of sarcomatoid squamous cell carcinoma (SpCC) remains unclear due to its rarity. Radiotherapy can be utilised as a standalone treatment; however, mesenchymal cells often show resistance to radiation, necessitating a combination with surgical interventions to minimise local recurrence [7]. In the present case, radical radiotherapy was administered for a T1 glottic carcinoma, resulting in an excellent response from the patient. The treatment approach reflects the need for tailored strategies, considering the aggressive nature of SpCC and the importance of comprehensive evaluation for effective management.

The squamous cell component typically shows expression of markers such as AE1/AE3, CK1, CK8, CK9, Epithelial Membrane Antigen (EMA), K18 and p63. In contrast, the spindle cell component generally expresses vimentin along with other mesenchymal markers [6]. Research has indicated that for SpCCs featuring poorly differentiated epithelial components, p53, an essential transcription factor for epithelial proliferation and differentiation, which serves as a valuable diagnostic tool for SpCC in the head and neck region [8]. Immunohistochemistry plays a vital role in distinguishing between these two types: SpCC is characterised by positivity for cytokeratin and vimentin while being negative for Anaplastic Lymphoma Kinase 1 (ALK-1) conversely, Inflammatory Myofibroblastic Tumours (IMT) are positive for ALK-1 and vimentin but negative for cytokeratin [9].

The objectives of treatment for SpCC include curing the cancer, preserving the larynx, ensuring good postoperative voice quality, and minimising complications [7]. Surgical excision with wide margins is the primary approach, as SpCC is known to have low sensitivity to radiation and chemotherapy [6]. In early stages, polypectomy and wide local excision can effectively remove the tumour without invading the underlying stroma [8]. Endoscopic microsurgery techniques, such as using a cold knife or Potassium Titanyl Phosphate (KTP) laser, have been documented as effective methods [10]. For T2 stage tumours, conservative management with irradiation can help maintain vocal function [8]. In one case, the KTP/532 laser was successfully employed to completely excise a recurrent tumour, followed by radiotherapy. For tumours classified as stage 3 or 4, treatment options include local resection, partial or total laryngectomy with or without lymph node dissection, followed by a combination of radiation and chemotherapy [8].

The prognosis for laryngeal SpCC is generally poorer than that for laryngeal SCC due to its highly malignant nature [6]. Factors influencing prognosis include tumour subsite, stage and size [6]. It has been indicated that laryngeal SpCC has a high rate of local recurrence, while regional lymph node involvement and distant metastases are relatively uncommon [6]. The five-year survival rate for patients with laryngeal SpCC ranges from 65% to 95% [7]. Most cases are diagnosed early, at stages T1 and T2, which correlates with a better prognosis. Favourable prognostic indicators include low tumour staging, glottic location, preserved vocal cord mobility, and no history of prior irradiation [9]. Conversely, poor prognostic factors encompass high tumour staging, large tumours (greater than 3 cm), predominance of the epithelial component, non glottic tumours, vocal cord fixation, previous radiotherapy and metastasis to regional or distant sites [8]. Additionally, lower levels of cytokeratin expression have been associated with improved survival rates [9]. Long-term follow-up after treatment is crucial for these patients.

CONCLUSION(S)

Laryngeal SpCC is a rare malignancy that presents significant diagnostic challenges in otolaryngology. Key factors aiding diagnosis include clinical appearance, age, gender and history of smoking or alcohol consumption. A confirmatory diagnosis requires histopathological analysis with immunohistochemistry, utilising both epithelial and mesenchymal markers. Treatment primarily involves surgical excision, while radiotherapy is indicated for recurrent or extensive cases; in rare instances, total laryngectomy may be necessary.

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