

Spindle cell carcinoma of the larynx: A rare case report

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ABSTRACT

Spindle cell carcinoma (SpCC) of the larynx, a subtype and a more aggressive variant of the commonly occurring squamous cell carcinoma, is a unique and rare neoplasm. It comprises of 0.6–1.5% of all laryngeal cancers. Macroscopically, it usually presents as a large pedunculated, polypoidal mass with surface ulceration. Microscopically, however, it is considered as a biphasic tumor that has surface epithelial changes (*in situ* to invasive carcinoma) and an underlying mesenchymal spindle shaped neoplastic proliferation. If detected early, it has a very good prognosis. We present a case of SpCC in a 70-year-old male, who presented with progressive hoarseness since 1 year. The mass was removed under videolaryngoscopic guidance and thereafter, the patient underwent cobalt 60 radiotherapy. His symptoms gradually improved, and he regained good control of his voice.

Key words: Biphasic, larynx, spindle cell carcinoma

INTRODUCTION

Spindle cell carcinoma (SpCC) designates a rare variant of squamous cell carcinoma (SCC) in which the spindle epithelial cell resembles a sarcoma on histological examination. It comprises 0.6–1.5% of all laryngeal cancers.^[1] It is also known as pseudosarcoma, carcinosarcoma, sarcomatoid SCC, or polypoid SCC. It usually presents as a large polypoid, pedunculated neoplasm, protruding from the mucosal surface with surface ulceration. SpCC is most commonly seen in the head and neck region. Most cases in the head and neck arise in the oral cavity, tonsil, pharynx, and larynx.^[2] Since a majority of these tumors are polypoid or pedunculated and tend to cause obstructive symptoms, these tumors are often detected at an early stage and tend to have a very good prognosis. The mean age of appearance for SpCC is the sixth decade of life, and it has a male predominance.^[3] The predisposing factors are the same

as SCC, including tobacco use, alcohol abuse, smoking, poor oral hygiene, and previous irradiation to the site. The following is a case report of one such patient, who presented to our institution with spindle cell (sarcomatoid) carcinoma of the larynx.

CASE REPORT

A 70-year-old male presented to our department with a 1-year history of progressive hoarseness and stridor since last 1 week. The stridor was biphasic in nature and had occurred after he had suffered an acute attack of upper respiratory tract infection. There were no associated complaints of dysphagia, odynophagia, or weight loss. There was no history of tuberculosis, trauma, or any focal neurological deficit. He was a chronic smoker since last 40 years.

On examination, the patient had no palpable neck nodes. Laryngeal crepitus was present. On flexible nasopharyngolaryngoscopy, a polypoidal, pedunculated

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mass with surface ulceration was seen arising from the right vocal cord, obstructing the glottic chink and extending into the subglottis [Figure 1].

The patient underwent tracheostomy under local anesthesia followed by videolaryngoscopy guided removal of the pedunculated mass under general anesthesia. The excised mass was sent for histopathological examination (HPE). The HPE report suggested spindle cell (sarcomatoid) carcinoma [Figures 2 and 3]. The patient was decannulated after 5 days and thereafter, he underwent cobalt 60 radiotherapy (30 exposures over 6 weeks period). He was followed up on a monthly basis for a period of 6 months. During this period his symptoms gradually improved, and he regained good control of his voice along with no recurrence.

DISCUSSION

SCC is considered to be the most common type of malignant laryngeal tumor.^[1] SpCC or sarcomatoid carcinoma is a highly malignant variant of it. Virchow was the first person to introduce the term. Numerous hypotheses regarding the histogenesis of SpCC have been proposed. Three dominant pathogenetic theories have been proposed: The tumor (1) represents a “collision tumor” (carcinosarcoma) (2) is an SCC with an atypical reactive stroma (pseudosarcoma) or (3) is of epithelial origin with transformation to spindle cell morphology (sarcomatoid carcinoma).^[3] Out of these, the third theory is the most accepted one now. Although the exact cause of SpCC is not known, it is strongly associated with a history of cigarette smoking and alcohol abuse. It has also been suggested that SpCC is associated with radiation exposure. SpCC is more predominant in males compared to females (12:1 ratio). SpCC most commonly affects the glottis in the majority of cases (70%), and the majority of patients present with symptoms of hoarseness, dyspnea, cough, and dysphagia often of <1-year duration.^[4] The majority of these tumors are characterized as being polypoid or pedunculated (98.9%) tumors that are often <2 cm.^[5]

Differential diagnosis includes a number of benign and malignant tumors, such as fibromatosis, nodular fasciitis, reactive epithelial proliferations, SCC, fibrosarcoma, malignant fibrous histiocytoma, leiomyosarcoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumor, mesenchymal chondrosarcoma, and malignant melanoma.^[6]

Tumors are staged according to their size and the presence or absence of metastasis to local lymph nodes or other parts of the body.^[3] Tumor size, location (supraglottic, glottis, or subglottic), vocal cord mobility, and tumor stage can all impact the treatment and outcomes of patients. The majority

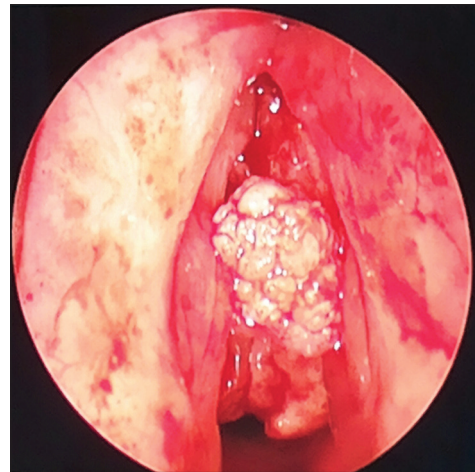


Figure 1: A polypoid, pedunculated mass with surface ulceration involving the right vocal cord, obstructing the glottic chink, and extending into subglottis

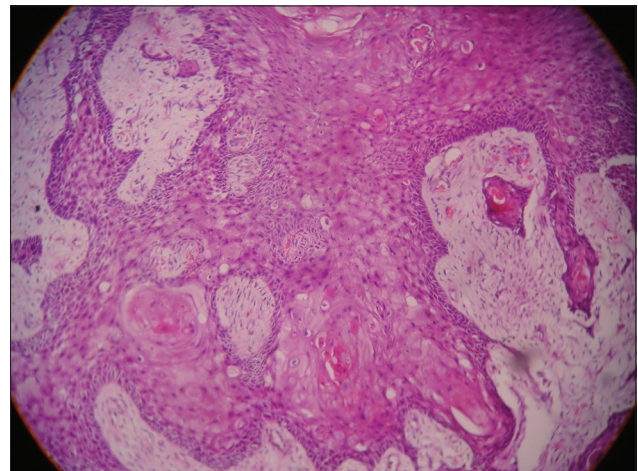


Figure 2: (H and E, ×100) section shows hyperplastic, irregular acanthotic squamous epithelium invading into fibrostromal tissue

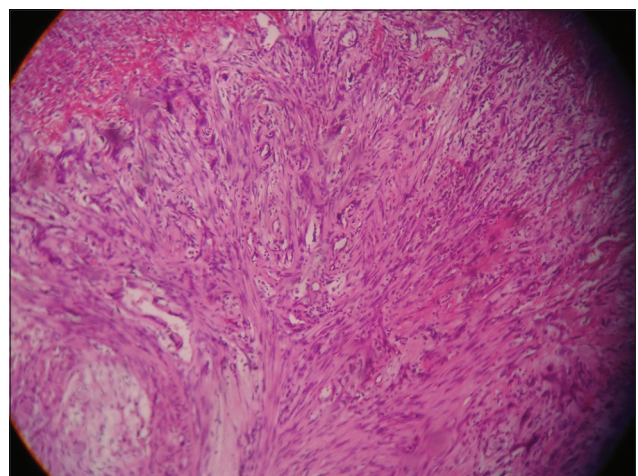


Figure 3: (H and E, ×100) section shows subepithelial proliferation of malignant spindle shaped cells arranged in haphazardly and fascicles

of spindle cell tumors are detected early in stages T1 and T2 due to their obstruction of the larynx causing symptoms and are correlated with a better prognosis.^[7]

It is axiomatic that the goals of treatment encompass cure of cancer, laryngeal preservation to the greatest degree possible, good voice quality posttherapeutically, and a low risk of complications. Since most spindle cell tumors are polypoid and pedunculated, polypectomy, and wide local excision can completely eliminate the entire tumor mass, because of their noninvasion of the underlying stroma at that early stage.^[8] Tumors that are stage 3–4 can be treated with local resection, partial laryngectomy, and total laryngectomy with or without lymph node dissection followed by a combination of radiation therapy and chemotherapy.^[4] SpCC of the larynx has a very good 5-year prognosis of 65–95%.^[11] Poor prognostic factors include tumors diagnosed at higher stages, large tumors (3 cm) with a predominance of epithelial component, nonglottic tumors, fixed vocal cords, history of radiotherapy and metastasis to regional lymph nodes, and distant metastasis.^[4]

CONCLUSION

According to the recent WHO definition, laryngeal SpCCs represent biphasic tumors, composed of an SCC, either *in situ* or invasive, and a malignant spindle cell component with mesenchymal appearance. It is a highly malignant variant of SCC that is, very uncommon. Because most spindle cell tumors are polypoid and pedunculated, and tend to cause obstructive symptoms such as hoarseness, dyspnea, and dysphagia, most tumors without metastasis are detected early and tend to have a very good 5-year prognosis.

This patient's SpCC was staged T1N0 without any evidence of metastasis. Due to the early detection of the tumor, the

patient underwent surgical excision and adjuvant radiation therapy. His symptoms subsequently improved, and he regained good control of his voice.

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Conflicts of interest

There are no conflicts of interest.

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