Case Report

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Carcinosarcoma (Spindle Cell Variant) of Larynx – Role of Radiotherapy

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Abstract

Carcinosarcoma, a highly malignant variant of squamous cell carcinoma (SCC) is uncommon, and it comprises of <2% of all laryngeal carcinomas. They are aggressive tumors but usually have a good prognosis since they are detected at an early stage due to obstructive symptoms. They are usually pedunculated or polypoidal in growth. Here, we present a case of a 63-year-old male patient who came with complaints of hoarseness of voice since 4 months and was diagnosed to be a case of spindle cell carcinoma of right vocal cord. The patient was treated with definitive radiation therapy and voice preservation was achieved.

Key words: Radiotherapy, Larynx, Carcinosarcoma

INTRODUCTION

Carcinosarcoma of the larynx is a highly malignant variant of squamous cell carcinoma (SCC). It is uncommon and comprises of <2% of all laryngeal carcinomas. They are biphasic in nature as they have surface epithelial changes and an underlying spindle shaped neoplastic proliferation. It is also known as pseudosarcoma, carcinosarcoma, sarcomatoid SCC, or polypoid SCC. They are tumors with aggressive potential. On direct laryngoscopy, they are detected as polypoidal or pedunculated mass. They are detected at an early stage due to obstructive symptoms such as dyspnea, odynophagia, progressive hoarseness of voice and hence have a good prognosis. The following is a case report of a patient who presented to our institution with carcinosarcoma (spindle cell variant) of right vocal cord.

CASE REPORT

A previously healthy 63-year-old elderly male patient presented with complaints of hoarseness of voice since 4 months, insidious onset, gradually progressive. The patient had a history of smoking 20 beedi/day for 4 months. He also gave a history of alcohol consumption for 40 years. There is no history of any medical co-morbidities.

On clinical examination, there was no evidence of cervical lymphadenopathy and on indirect laryngoscopy - smooth pale exophytic mass seen involving right vocal cord and also anterior commissure with edema of the left false vocal cord/ventricle. Right vocal cord restricted mobility.

CT scan head and neck showed enhancing lesion noted in the right glottis measuring 10 * 6 mm protruding into the lumen(Figure 1). Paraglottic space appears normal. No evidence of infiltration of the adjacent structures or cervical lymphadenopathy.

Direct laryngoscopy with biopsy - growth noted over the right false cord, AE fold, true cord, with subglottis extension. Impaired vocal cord mobility was seen.

Histopathology report shows fragments of mucosal tissue with tumor seen (Figure 2). The superficial squamous epithelium is seen focally with areas of denudation. A spindle cell tumor composed of spindle cells arranged in short and long fascicles with storiform pattern seen. A focus of necrosis seen. Features suggestive of spindle cell carcinoma:

- Immunohistochemistry (IHC): Strongly positive for CK and S100 and negative for DESMIN, smooth muscle actin (SMA), and CD34.
- Chest X-Ray: Normal.

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The patient was staged as T2N0M0, as the tumor had subglottic extension with impaired vocal cord mobility without any cervical lymphadenopathy or distant metastasis. The patient was not amenable for surgery due to subglottic extension and was planned for definitive radiation therapy and received 6000 cGy in 25 fractions. On follow-up, the patient was asymptomatic and was assessed radiologically, which showed no evidence of tumor and voice preservation was achieved. (Figure 3)

DISCUSSION

SCC is considered to be the most common type of malignant laryngeal tumor. Carcinosarcoma of the larynx is a highly malignant variant of SCC. It is a rare tumor, which comprises around 2% of all laryngeal cancers.[1] Spindle cell carcinoma is also called as biphasic tumor because it has squamous cell (*in situ* or invasive) with a spindle cell stromal reactions.[2] SpCC is also considered to be a monoclonal epithelial neoplasm with the sarcomatous component derived from squamous epithelium with divergent mesenchymal differentiation.[2] It is also known as pseudosarcoma, carcinosarcoma, sarcomatoid SCC, or polypoid SCC. It can arise from any site on the body, but the most common site of origin is head and neck region mainly oral cavity, tonsil, larynx, and pharynx. Exact etiology of SpCC is unknown; however, it is associated with a history of chronic cigarette smoking and alcohol abuse.[3] SpCC has peak incidence between 6th and 7th decades and has male preponderance when compared to females (12:1).[4]

The most common presenting symptoms will be hoarseness, dyspnea, cough, dysphagia, and odynophagia.[1] These tumors are usually polypoid or pedunculated masses.[4] Spindle cell carcinoma is mainly diagnosed pathologically in which both squamous cell and a spindle cell stromal reaction (sarcomatous) histology is demonstrated.[2] Here, in our case, fragments of mucosal tissue with the tumor were seen, and superficial squamous epithelium was seen focally with areas of denudation. A spindle cell tumor composed of spindle cells arranged in short and long fascicles with storiform pattern and a focus of necrosis seen with 10-12 Mitosis per 10 hpf which was suggestive of spindle cell carcinoma. Further, immunohistochemical studies of epithelial and mesenchymal markers are used to diagnose the tumor. Epithelial markers include keratin (AE1/AE3, CK1, 8, 9), epithelial membrane antigens, KI, and K18. Mesenchymal markers include vimentin, DESMIN, S-100, osteopontin, and bone morphogenetic protein (2, 4).[5]

In our case, IHC was strongly positive for CK and S100 and negative for DESMIN, SMA, and CD34 leading to a diagnosis of spindle cell carcinoma. Management is mainly based on the tumor site and T stage of the tumor. Since most of the cases are diagnosed in early stages due to their obstructive symptoms, they can be managed surgically

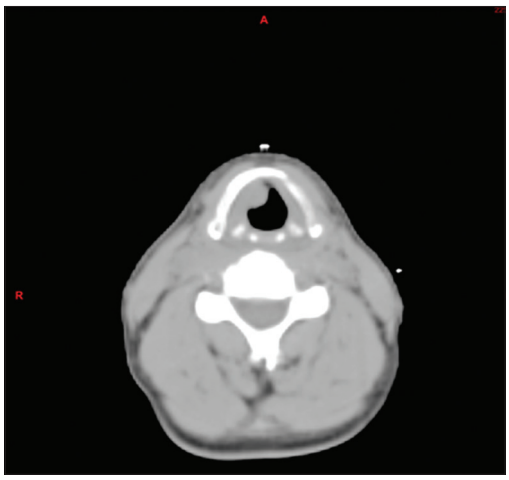

Figure 1: CT scan head and neck showed enhancing lesion noted in the right glottis measuring 10 * 6 mm protruding into lumen. However, paraglottic space appears normal. No evidence of infiltration of the adjacent structures or cervical lymphadenopathy

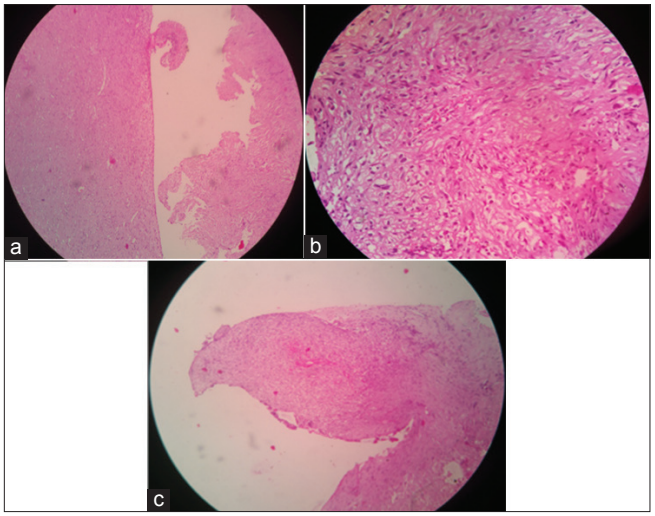

Figure 2: (a-c) Histopathology report shows fragments of mucosal tissue with tumor seen. Superficial squamous epithelium is seen focally with areas of denudation. A spindle cell tumor composed of spindle cells arranged in short and long fascicles with storiform pattern seen. A focus of necrosis seen. Features suggestive of spindle cell carcinoma

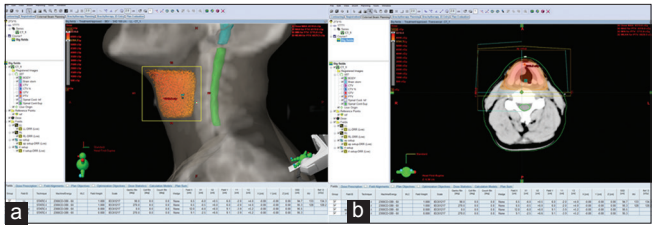

Figure 3: (a-b) Radiotherapy portals in dose color wash with 95% dose coverage in: (a) Lateral and (b) Axial view

followed by radiation. Chemotherapy is usually not preferred in sarcomas as they are chemoresistant. Surgery was deferred in our case due to subglottic extension and in view of voice preservation and was planned for definitive radiation.

In previous studies, local recurrence rate has been reported between 16% and 32%.[1] Cervical nodal metastasis is reported between 7% and 26%.[4,6,7] Lambert *et al.* reported 5% of distant metastasis to the lung and soft tissue.[8] Carcinosarcoma of the larynx has a very good 5-year prognosis of 65-95%.[9]

CONCLUSION

Carcinosarcoma of the larynx is a highly malignant variant of SCC. They are tumors with aggressive potential. They are detected at an early stage due to obstructive symptoms such as dyspnea, odynophagia, progressive hoarseness of voice and hence have a good prognosis.

This patient was staged as T2 N0 M0 and in view of subglottic extension surgery was deferred, and the patient received definitive radiotherapy. Hence, definitive radiotherapy can be used as an alternative management in cases whom surgery is deferred with good voice preservation.

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