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Case Report

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Sarcomatoid carcinoma a rare variant of squamous cell carcinoma of larynx: a case report

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ABSTRACT

Sarcomatoid (spindle cell) carcinoma is a rare variant of squamous cell carcinoma (SCC). It compromises of 2–3% of laryngeal cancers. Tumor arises from the oral cavity, tonsil, larynx and pharynx. Majority of these tumors are polypoid or pedunculated and tend to cause obstructive symptoms. These tumors are often detected at an early stage, removed by polypectomy during diagnosis and tend to have a very good prognosis. In this case report, 76 years male who presented with progressive hoarseness of his voice and pain in throat. The patient underwent direct laryngoscopy with excision of the malignant mass and received radiotherapy.

Keywords: Sarcomatoid, Pedunculated, Polypectomy, Laryngoscopy

INTRODUCTION

Squamous cell carcinoma is the most malignant carcinoma of the larynx.[1] Sarcomatoid tumor is considered a highly malignant variant of squamous cell carcinoma and rare tumor of larynx. It is a unique and rare neoplasm, composed of both malignant epithelial and mesenchymal components. Till date, only few cases are reported of sarcomatoid carcinoma of 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 following is a case report of one such patient who presented to our institution with sarcomatoid carcinoma of the larynx.

CASE REPORT

A 76 years old male presented to the ENT OPD with 3 months history of progressive hoarseness and pain in throat. The patient history was significant for 40 pack years of smoking. There was no history of hemoptysis, weight loss, neck masses or any signs suggestive of any chronic debilitating disease. Physical examination of the patient showed no palpable cervical lymphadenopathy.

The patient underwent a flexible laryngoscopy that showed the presence of whitish polypoidal mass involving left true vocal cord (Figure 1). The patient then underwent direct laryngoscopy, which showed a mass with filiform projections located in the midregion of left true vocal cord. The lesion was excised and sent for histopathological examination.

The pathology report showed a sarcomatoid carcinoma (Figure 2) that on immunohistochemistry strongly positive for Ki -67, SMA and focal positive for EMA but negative for desmin, HMB – 45, CK 5/6, CK HMW, Anti p63 and S-100.

A CT scan of his neck with contrast after the histopathological confirmation revealed a minimal thickening of size 1×0.5 cm involving the left true vocal cord (Figure 3); however the nasopharynx, pharynx, parapharyngeal space, epiglottis, major neck vessels appeared normal. The parotid glands and submandibular

salivary glands were normal. There was no significant cervical lymphadenopathy.

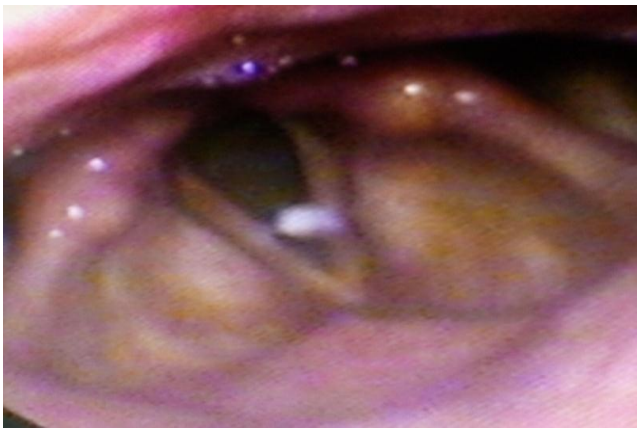

Figure 1: Fiberoptic laryngoscopy showing whitish polypoidal mass involving the left true vocal cords.

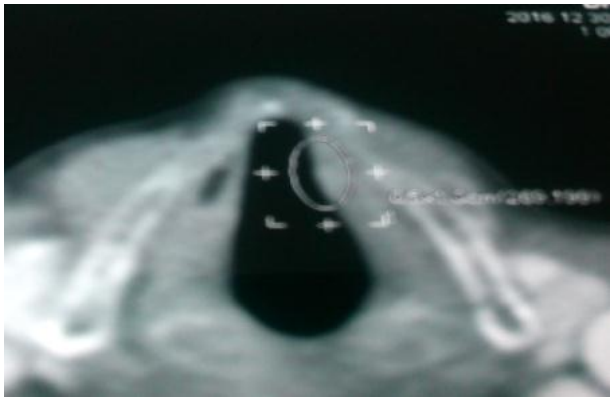

Figure 2: Computed tomography neck showing minimal thickening of size 5×1 cm involving the left true vocal cord.

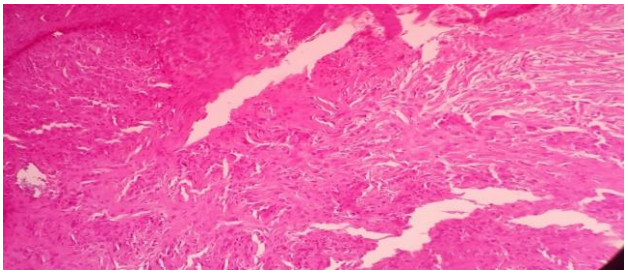

Figure 3: Histological findings.

Lining epithelium shows acanthosis and parakeratosis. Stroma shows pleomorphic spindle cell in isolation and in fascicles. Spindle cells have hyperchromatic, pleomorphic nuclei with eosinophilic cytoplasm. Occasional cells show intranuclear inclusions and occasional mitotic figures. Occasional bi, trinucleate and pleomorphic giant cells are seen. Focal areas show dense collagen bundles. Histopathology revealed sarcomatoid carcinoma variant of squamous cell carcinoma.

The patient sarcomatoid carcinoma was staged as T1N0M0 after investigations and received radiotherapy. The patient's symptoms gradually improved and complete resolution of tumor after radiotherapy (Figure 4).

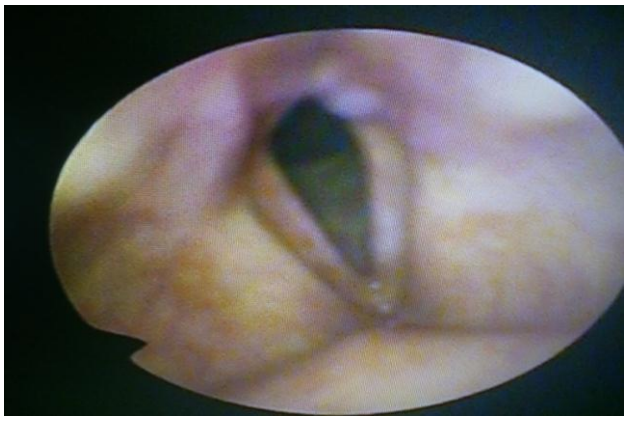

Figure 4: After radiotherapy fiberoptic laryngoscopy showing complete resolution of tumour.

DISCUSSION

SCC is considered to be the most common type of malignant laryngeal tumor.[1] Sarcomatoid carcinoma is a highly malignant variant of SCC. It is a rare tumor with a reported incidence of 2–3% of all laryngeal cancers.[2] Sarcomatoid is an aggressive malignant tumor typically observed in upper aerodigestive tract, in which nearly half of the cases are of larynx origin.[3] It was first reported in the literature in 1933 by Figi, under the name of "larynx sarcoma".[4]

Although the exact cause of spindle cell carcinoma (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 although the determination of radiation risk may be complicated by the dose and duration of radiation exposure.[5] SpCC is more predominant in men compared to females (12:1 ratio) although it is becoming more common in females and it is usually seen in the 6th and 7th decades of life.[6]

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.[1] The majority of these tumors are characterized as being polypoid or pedunculated (98.9%) tumors that are often <2 cm in size.[6]

The diagnosis of SpCC requires histological demonstration of both the squamous cell component and the spindle shape cells with sarcomatous appearances.[2] The histological examination can often show the presence of SCC at the surface or deeper within the tumor although this is rare especially with tumors where the surface is ulcerated or denuded. It is often seen is a blending of squamous cells and spindle cells which can be differentiated by their different arrangement which includes storiform, solid, and fascicular appearance.[6]

In addition to histological studies, 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 BMP (2, 4).[7] For spindle cell carcinomas with poorly differentiated epithelial tumor components Lewis et al. have shown that p53, a transcription factor that is important for epithelial proliferation and differentiation, is particularly useful for diagnosing SpCC of the head and neck region.[8]

Dubal et al reported that the majority of laryngeal SpCC cases have T1 (62%) and stage I (61%) tumors at presentation.[9] Lymph node metastasis may occur (12.6%) but distant metastasis is uncommon (3.7%). he surgery yields a favorable prognosis when compared to radiotherapy. The 5-year disease speciic survival (DSS) is 84% with surgery alone, 84.2% with surgery combined with radiotherapy, and 60.5% with radiotherapy alone. In addition, a glottis location of the tumor, as opposed to a non-glottic location, is significantly associated with improved prognosis (5 year DSS, 84.0% versus 51.9%). Compared to other laryngeal malignancies, SpCCs have intermediate (5 years) survival benefit (74.1% versus 64.6%), but short-term (1 year, 90.9% versus 88%) and long-term (10 years, 57.9% versus 50.6%) survival rates are similar.

On the other hand, in a retrospective study assessing the outcomes of early-stage (T1-T2) glottic SpCC cases (n =28) treated with primary radiotherapy. Ballo et al reported that these patients had similar local-control rates to irradiated patients with similar volume disease with the more typical squamous cell carcinoma.[10] Therefore, the authors suggested that the histologic diagnosis of SpCC instead of squamous cell carcinoma should not alter treatment recommendations in patients with early-stage disease.

Tumors that are stage T2 or less can be managed conservatively with limited field irradiation and conservative treatment to preserve the patient's voice. Tumors that are stage 3–4 can be treated with local resection, partial laryngectomy, total laryngectomy with or without lymph node dissection followed by a combination of radiation therapy and chemotherapy.[6] SpCC of the larynx has a very good 5-year prognosis of 65–95%.[1] 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.[6]

CONCLUSION

SpCC or sarcomatoid carcinoma of the larynx 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.

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