



CARCINOSARCOMA OF LARYNX: A RARE CASE WITH EXCELLENT OUTCOME WITH STANDARD TREATMENT

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Article Received on 30/09/2015

Article Revised on 24/10/2015

Article Accepted on 18/11/2015

ABSTRACT

We report a case of a 60-year-old previously healthy male with carcinosarcoma of larynx who presented with hoarseness of voice. Carcinosarcoma carcinoma mimics sarcoma and carcinoma in histopathological parameters and it is difficult to categorize the histopathological type of sarcoma which is usually supplemented by immunohistochemistry for various markers. Carcinosarcoma of larynx is an uncommon presentation. The standard treatment is surgery and/or adjuvant radical external beam radiotherapy with standard fractionation schedule. Surgery is usually done in early lesions and reserved for advanced lesions. Advanced lesions are treated by radiotherapy alone as primary treatment. The prognosis of carcinosarcoma of larynx is felt to be extremely poor with high recurrence rate. Contrary to it, this case demonstrates that a good response with standard therapy is possible.

KEYWORDS: Carcinosarcoma, surgery, radiotherapy, larynx.

INTRODUCTION

Carcinosarcoma is an uncommon form of cancer with histological, cytological, or molecular properties of both mesenchymal tumors ("sarcoma") and epithelial tumors ("carcinoma").^[1] Laryngeal cancer presents 2% of the total cancer risk. Carcinosarcoma accounts for less than 1% of head and neck cancers.^[2] Male to female ratio is 11:1. This tumor usually develops in the 7th decade of life (mean age is 65.6 years). Possible etiologies are radiation, tobacco use and/or alcohol consumption.¹ Hoarseness is the chief complaint, accompanied by other nonspecific symptoms usually of short duration (<1 year) before directing the patient to seek medical attention.^[3] Despite to the radical treatment in the form of surgery and/or radiation therapy recurrence rate is high in the range of 45%. We report a case of 60-year-male who presented with hoarseness of voice and excellent prognosis to radical treatment without any recurrence.

CASE SUMMARY

A 60-year-male presented with hoarseness of voice of six months duration. It was insidious in onset and gradually progressive in nature. There were no aggravating or relieving factors and no associated history of throat pain, difficulty in breathing or difficulty in swallowing. No other significant comorbidities were noted. Patient was non smoker and non alcoholic. Complete hemogram and

all routine blood biochemistry parameters of the patients were within normal limits. A radiograph of chest did not reveal any metastatic nodules. Ultrasonography of the abdomen and pelvis region of the patient was normal. General physical and systemic examination of the patient was normal. Indirect laryngoscopy revealed growth on left vocal cord extending to anterior commissure and bilateral vocal cords were mobile. There was no significant lymphadenopathy associated with it. Contrast enhanced computed tomography scan revealed heterogeneously enhancing lesion of size 1.8 x 1.5 x 1.1 cm involving anterior commissure of larynx. The lesion was causing involvement of bilateral true vocal cords anteriorly and extending inferiorly into subglottic region along with partial obstruction of laryngeal airway. Few subcentimetric homogenous lymph nodes were seen in bilateral jugular and posterior cervical region. Histopathological examination of the biopsy taken from the growth vocal cord revealed pleomorphic malignant tumor of larynx. On immunohistochemistry, the results were strongly positive for vimentin, focally positive for CD68, S-100 and CK, positive for CD31 and CD34 and negative for desmin, HMB 45, and EMA. On the basis of morphological and immunohistochemistry findings, histopathological features favour poorly differentiated sarcomatoid carcinoma, a malignant mesenchymal tumor. Patient was clinically diagnosed as T₂N₀M₀, stage

II. With this diagnosis, patient underwent microlaryngeal surgery followed by adjuvant radical external beam radiotherapy for 64 Gy in 32 fractions in 6.2 weeks with 2 Gy per fraction for five days per week, on Tele-cobalt machine in supine position by bilateral parallel opposed fields to face and neck. Radiotherapy was tolerated well and patient had an uneventful post-radiation phase. Patient is asymptomatic and having complete response. Patient is without any evidence of tumor recurrence after six months of follow-up.

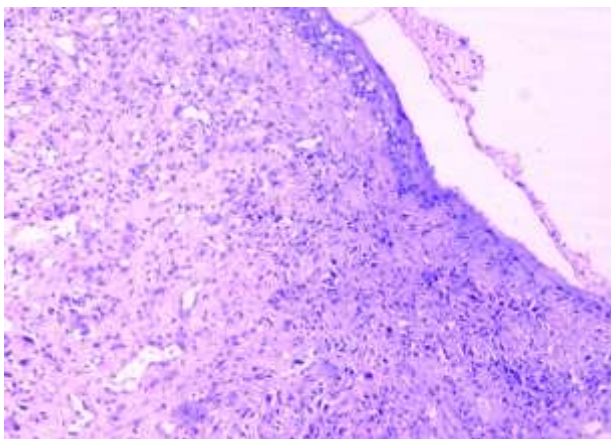


Figure 1: Biopsy revealing fibrocollagenous tissue infiltrated by singly scattered tumor cells, proliferating blood vessels, areas of necrosis alongwith mixed inflammatory infiltrate. (H&E 40x)

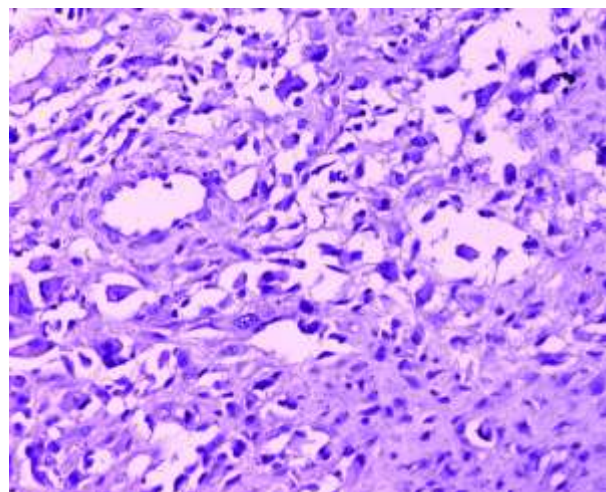


Figure 2: Oval to polygonal shaped tumor cells having hyperchromatic nuclei, variable cytoplasm alongwith blood vessels and tumor giant cells. (H&E 400x)

DISCUSSION

Carcinosarcomas are uncommon tumors frequently misdiagnosed as mesenchymal malignancies.^[4] Despite several electron microscopic, immunohistochemical and genetic studies, precise histogenesis of carcinosarcoma is quite controversial.^[9] It is also known as “sarcomatoid carcinoma,” “spindle cell carcinoma,” “pseudosarcoma,” “spindle squamous cell carcinoma,” “collision tumor,” or “Lane tumor”.^[4] The term ‘spindle cell carcinoma’ was

first applied by Shervin et al.^[8] Carcinosarcoma of larynx accounts for 1-2% of all primary benign and malignant laryngeal neoplasms and 4.2% of squamous cell carcinoma of the larynx. 7.7% of the cases are associated with previous exposure to radiation.^[1] Most common site of involvement is vocal fold.^[4] Most of the lesions are polypoidal in nature, others may be ulcerated or sessile^[5] and these tend to obstruct the larynx and cause symptoms in early stages of the disease.^[1] Glottis tumors are the most common constituting around 70%, left sided lesions are more common. Most of these present in early stage, T1 or T2.^[1,6] As presented in our case, hoarseness is the most frequent symptom^[3], present in around 88% of the patients. Other symptoms are change of voice, breathlessness, airway obstruction, difficulty in swallowing, sore throat, cough or stridor.^[6] Patients with previous history of radiation exposure and women may present with shorter duration of symptoms.¹ In vocal cord carcinoma, at diagnosis, the incidence of clinically positive lymph nodes approaches zero for T₁ lesions and less than 2% for T₂ lesions. The incidence of neck metastases increases to 20% to 30% for T₃ and T₄ lesions.^[2] Distant metastasis occurs in 5-15%.^[4] Distant metastasis are most common in lung and less common in cervical lymph nodes, liver, kidney, pleura, brain or bone (jaw and vertebrae).^[3] Metastasis is more commonly seen with subglottic and supraglottic tumors than with glottic tumors.^[1] Carcinosarcoma of the larynx has a similar reputation for radioresistance and for dedifferentiation after RT. The 5-year actuarial control rate is 89%, after irradiating T₁₋₂ glottic carcinosarcoma, which is similar to that obtained for squamous cell carcinoma.¹²⁵ All patients with recurrences could undergo salvage surgery.^[3]

Contrast enhanced CT scan is the investigation of choice for studying larynx. Although minimal mucosal lesions are not detected by CT scan.^[2] MRI is used to define early cartilage destruction or subtle exolaryngeal spread.^[2] Vimentin and keratin antibodies may contribute to detect distant metastasis.^[6] AJCC 2010 staging system is used for staging of laryngeal cancer.² Differential diagnosis include a number of benign and malignant processes, such as fibrosarcoma, leiomyosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, malignant peripheral nerve sheath tumor, osteosarcoma, synovial sarcoma, malignant melanoma, mesenchymal chondrosarcoma, Kaposi's sarcoma, angiosarcoma, leiomyoma, fibromatosis, inflammatory myofibroblastic tumor, nodular fasciitis or reactive epithelial proliferations.^[1,3,4]

Standard treatment is surgery alone or surgery followed by adjuvant radiation therapy.^[1] Irradiation is the initial treatment for T1 and T2 lesions, and surgery is reserved for salvage after failure of radiation therapy.^[2] The various surgical modalities include vocal cord stripping, partial laryngectomy, hemilaryngectomy, supraglottic laryngectomy, microlaryngeal surgery, or laryngectomy, with or without lymph node dissection (partial, modified or radical neck dissection). Radiation treatment involves

external beam irradiation to the larynx and neck. The major advantage of irradiation compared with partial laryngectomy is organ preservation and better quality of life.^[1,3] Prognosis of carcinosarcoma larynx is better than classic squamous cell carcinoma of larynx.^[6] The 5-year actuarial local control rates are 94% and 54% for patients with T1 and T2 lesions respectively. The 10-year actuarial disease-specific and overall survival rates are 92% and 63%, respectively. Five year overall survival rate is 58.8%. Recurrence rate is 45%, and it may occur from within 2 months to 9 years post treatment. More favourable prognosis is associated with glottis tumors. Local control in early T₁ or T₂ glottic tumors is 70-96% with radiation alone and 63-100% with surgery alone.^[1]

In the present case, patient underwent microlaryngeal surgery of larynx followed by adjuvant radical radiation therapy. After completing radiation therapy successfully, patient is having complete response and without any evidence of tumor recurrence even after six months of follow up.^[1]

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