SARCOMATOID CARCINOMA OF THE OESOPHAGUS: A DIAGNOSTIC DILEMMA, A MENACE TO TREAT!

Junaid M, Armed Forces Hospital Southern Region
Khamis Mushayt, Saudia Arabia
Qadeer S, Sir Syed College of Medical Sciences for Girls, Pakistan
Al Syed G, Armed Forces Hospital Southern Region
Khamis Mushayt, Saudia Arabia

Corresponding author: Junaid M, Surgery Consultant-Otolaryngology-head and neck surgery, Armed Forces Hospital Southern Region Khamis Mushayt, Saudia Arabia. E-mail: montsj@gmail.com

Citation: Junaid, M., Qadeer, S., & Al Syed, G. (2019). Sarcomatoid Carcinoma of the Oesophagus: A Diagnostic Dilemma, a Menace to Treat!. Frontiers Journal of Case Reports and Images, 1(1), 1-5.

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Received Date: 30 July 2019; Accepted Date: 16 August 2019; Published Date: 23 August 2019.

ABSTRACT

Sarcomatoid carcinoma may be a rare entity in head and neck but is a potentially aggressive tumor with a tough diagnostic dilemma. It has a tendency to recur and metastasize easily and efforts should be made to control loco-regional and distant spread. Study of more cases will help in assessing its behaviour and decide on optimal treatment options for this aggressive tumor.

Keywords: Sarcomatoid carcinoma, Pseudosarcoma, Carcinosarcoma, Esophageal cancer, Neck swelling.

INTRODUCTION

Sarcomatoid carcinoma is referred as a variety of terms in literature. Names like carcinosarcoma, pseudosarcoma, sarcomatoid squamous cell carcinoma, pleomorphic carcinoma and most famously spindle cell carcinoma, all refer to the same entity (Roy et al.,
Within the head and neck, it is a rare malignancy, with the most frequent site to be effected is the larynx. It is also reported to be found in the nasal cavity, hypopharynx, oral cavities, oesophagus, trachea and skin. It can be defined as a poorly differentiated form of squamous cell carcinoma, with elongated epithelial cells, resembling sarcoma (Su et al., 2006). Discussed below is a case we came across at our institute and its management.

**CASE PRESENTATION**

Our patient was a seventy five years old lady, who developed gradual left sided neck swelling over a period of four years. Despite the fact that it continued to increase in size, the patient initially chose to ignore, as it was symptomless. About a year back, it became associated with hoarseness and progressive dysphagia. She also had intermittent breathing difficulties, most significant on lying down. She underwent biopsy of the neck mass at a private institute which showed malignant fibrous histiocytoma of neck. On examination, the swelling appeared on left side of the neck, in the midline. It measured about 14 cm x 12 cm x 10 cm and was hard, non-mobile or tender and non-fluctuant. Trachea was shifted to the contralateral side while there was an old surgical mark on the left side of the neck (level III-previous biopsy mark) and indirect laryngoscopy for visualisation of the vocal cords was unsuccessful (Figure 1).

![Figure 1](image)

**Figure 1**

Coronal view of a heterogenous mass with lateral deviation of the great vessels

A Pan computed tomography revealed a heterogeneous, well-defined mass in the left side of the neck, abutting the great vessels laterally. Posteriorly, it was extending into the prevertebral space and medially was displacing the thyroid cartilage, larynx and trachea towards the opposite side. There was obvious infra-clavicular extension as well (Figure 2). While chest abdomen and pelvis was found to be free of metastasis. An informed consent for high risk surgery was obtained. The patient was successfully intubated using a small lumen endotracheal tube. The mass was appreciated deep to the left sternomastoid (Figure 3). Dissection was started from the lateral side of the mass. The laryngeal apparatus and trachea though adherent to the mass, was separated and found to be not involved. However, the oesophageal wall was involved in the anterolateral aspect and had to be dissected out, creating a 3 cm defect which was primarily repaired. There was a large, adherent lymph node (> 3 cm) to the mass, at level V on left side and hence ipsilateral extended neck (I-VI) dissection was also performed. The left Sternocleidomastoid muscle, Jugular vein, left lobe of thyroid and strap muscles appeared to be involved with the disease, and subsequently removed while Carotid vagus nerve was preserved (Figure 4). The wound was closed over a low-suction drain.
FIGURE 2
Axial view of the mass showing significant deviation of the airway to the contralateral side

FIGURE 3
Mass behind the left sternomastoid occupying a large part of the neck

FIGURE 4
Carotid and Vagus nerve preserved
Post-operatively, the patient was kept intubated in ICU and extubated on 2nd post-operative day. She was shifted to a regular bed 2 days later and was discharged subsequently on 9th post-operative day. The final histopathology came out as sarcomatoid carcinoma of the oesophagus. Immuno-histo-chemistry showed positivity for cytokeratin and EMA while S100 and desmin were negative for the tumor cells. A week later, on follow up the wound was found to be healthy and clean. 4 weeks later, she was allowed to travel back to her city and referred to medical oncologist at the same time for appropriate therapy. Patient was contacted for follow-up 8 weeks, where she was found to have expired due to cardio pulmonary arrest at home.

**DISCUSSION**

Sarcomatoid carcinoma is an uncommon biphasic tumor with an even rarer occurrence in the head and neck. Within the head and neck, it is most frequently found in the larynx, pharynx, tonsillar fossa and the oral cavity where the incidence is less than 1% (Jordan & Regezi, 2003). The most frequent site of esophageal sarcomatoid is the distal end of the esophagus (60%), with the proximal part constituting only 10% of the tumors (Raza & Mazzara, 2011).

Poor oral hygiene, alcohol or tobacco intake as well as previous radiation are considered as risk factors for its development, as well as genetic predisposition (Takata et al., 1991). Predominantly found in males, it occurs generally in the seventh decade of life. They usually arise as painless exophytic, polypoidal lesions, with extensive surface ulceration. Morphologically, it has two components: the spindle cell or sarcomatoid component which makes up the bulk of the tumor and the epithelial component constituting the minor portion of the tumor. The most widely accepted theory for its histo-genetic nature is the monoclonal hypothesis stating that they originate from the same stem cells and ‘dedifferentiation’ has occurred (Miyajima et al., 2006).

It is extremely difficult and inconclusive to diagnose sarcomatoid carcinoma on light microscopy only. Sarcomatoid carcinoma shows positivity for EMA and hence, can be differentiated from other sarcomatous lesions using immuno-histochemical stains (Takata et al., 1991; Minami et al., 2008). Treatment is very similar to squamous cell carcinoma of the same stage. It is dependent on the site, extent, staging and metastasis of the tumor. Surgery is a widely accepted treatment option offered for oral and laryngeal tumors, with a better prognosis when compared with radiotherapy alone (Minami et al., 2008). Surgery is usually followed by radiation therapy or chemotherapy. Using radiation therapy as the primary treatment modality is not a favoured option any longer. Thomson et al, in his study, reported a better outcome for patients treated with surgery in comparison to radiation therapy treated patients (Romanach et al., 2010; Thompson et al., 2002). Chemotherapy as a treatment modality has ambiguous results (Su et al., 2006).

In comparison to squamous cell carcinoma, within the head and neck region, sarcomatoid carcinoma is considered as a more aggressive tumor, with a tendency to recur and metastasize early (Minami et al., 2008; Romanach et al., 2010). This in turn leads to poor prognosis. The presence of distant metastasis and the depth of invasion of the tumor are considered to be reliable prognostic features for the disease.

The involvement of cervical lymph nodes ranges between 7.5-26%, with an even lesser distant metastasis, mostly to the lungs (Thompson et al., 2002). Lambert et al in his study reported the incidence to be about 5% with the overall mortality rate reported in literature to be 14-32% (Thompson et al., 2002; Lambert et al., 1980).
CONCLUSION

Sarcomatoid carcinoma may be a rare entity in head and neck but is a potentially aggressive tumor with a tough diagnostic dilemma. It has a tendency to recur and metastasize easily and efforts should be made to control loco-regional and distant spread. Study of more cases will help in assessing its behaviour and decide on optimal treatment options for this aggressive tumor.

AUTHOR CONTRIBUTION

MJ is the primary surgeon, SQ performed the literature search, GS wrote the primary manuscript, MJ SQ proof read and arranged the figures and finalizes the manuscript.

REFERENCES


