



Sarcomatoid Carcinoma of the Pyriform Sinus: A Rare Malignancy in a Rare Location Responsive to Combination of Chemotherapy and Radiotherapy

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Sarcomatoid carcinoma (SaC) is a rare variant of squamous cell carcinoma with sarcomatoid features having biphasic characteristic malignant mesenchymal spindle cell and squamous cell component comprising of dysplasia, carcinoma *in situ*, or invasive carcinoma. Most information about this malignant entity in the head-and-neck region has emerged from larynx and oral cavity with an incidence of 0.5%–3% and 1%, respectively, with very few cases reported in hypopharynx. We hereby report such a case in a 76-year-old male who presented with dysphagia and swelling right side neck. Clinicopathological evaluation showed a localized SaC hypopharynx. In view of old age and comorbidities of hypertension and diabetes, no surgical intervention was contemplated, and he was treated with definitive concurrent chemo-radiotherapy (CCRT). A thorough review of the literature revealed that while earlier reported cases underwent upfront surgery followed by adjuvant RT or adjuvant CCRT, we treated this case with combination of chemotherapy and RT resulting in a significant therapeutic effect.

Key words: Sarcomatoid carcinoma, hypopharynx, pyriform sinus, chemotherapy, radiotherapy

INTRODUCTION

Sarcomatoid carcinoma (SaC) is a rare malignancy with unique biphasic histologic pattern reported in various organ systems¹ apart from head-and-neck (HN) with larynx being the most common location² with an incidence of 0.5%–3%^{4,5} followed by oral cavity with 1%.⁶ HN SaC is associated with a dismal prognosis as compared to extra-oral sites with higher recurrence and mortality due to extensive lymphatic and distant dissemination.¹ Surgery has been considered the standard of care for HN SaC⁴ but concurrent chemoradiotherapy (CCRT) as a definitive therapeutic modality may result in excellent clinical and radiological response, as seen in our case.

CASE REPORT

A 76-year-old male, chronic smoker, hypertensive and diabetic presented with dysphagia and swelling right side neck. Clinical examination showed conglomerate lymph node (LN) mass right neck cervical level II/III. Otorhinolaryngologic

examination showed a 3.8 cm × 2.5 cm growth right pyriform sinus (PFS), biopsy from which revealed pleomorphic spindle cell tumor with large hyperchromatic nuclei, prominent nucleoli with frequent mitosis with a mitotic rate of 4 per high-power-field arranged in intersecting fascicles. Few strips of squamous mucosa showing full thickness dysplasia were also present [Figure 1]. Immunohistochemistry (IHC)-stained positive for pan-cytokeratin (pan-CK) [Figure 2], p63 [Figure 3] and S-100 [Figure 4] while stained negative for smooth muscle actin (SMA) which gave a diagnosis of SaC of PFS.

Whole body 18-fluorodeoxyglucose positron emission computed tomography (WB-18-FDG-PET/CT) scan showed a 2.4 cm × 1.1 cm × 1.1 cm growth right PFS [Figure 5] with a standard uptake value (SUV) of 10.36 and LNs 6.2 cm × 4.2 cm right level II and 3.5 cm × 2.6 cm right level III with SUV of 19.99 and 8.81, respectively [Figure 6]. Given old age, multiple

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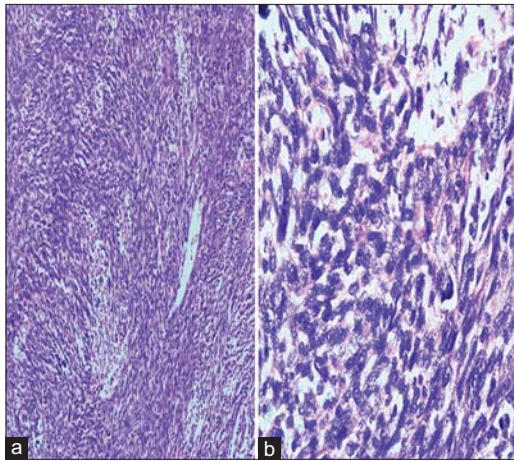


Figure 1: Biopsy from right pyriform sinus showing pleomorphic spindle cell tumor with large hyperchromatic nuclei, prominent nucleoli with frequent mitosis with a mitotic rate of 4 per high-power-field arranged in intersecting fascicles. Few strips of squamous mucosa showing full thickness dysplasia was also present (a) H and E, $\times 10$; (b) H and E, $\times 40$

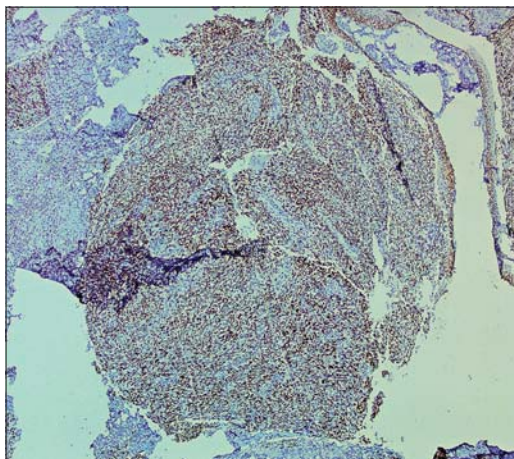


Figure 3: Immunohistochemistry staining positive for p63 ($\times 10$)

comorbidities and N3 nodal status with fixity to underlying structures indicating an advanced stage, no active surgical intervention was contemplated. He was treated with external beam RT to a dose of 70 Gy in 35 fractions along with 5 cycles of 50 mg weekly cisplatin given on a concurrent basis at 40 mg/m². Due to persistent nausea, vomiting, and leukopenia, the patient was not given the sixth chemotherapy cycle and also no further adjuvant chemotherapy was planned or given. He showed significant therapeutic benefit to treatment with improvement in dysphagia and complete resolution of the LN mass [Figure 7]. Clinical evaluation and WB-18-FDG-PET/CT 3 months post-CCRT showed complete resolution of the PFS growth along with cervical LNs [Figure 8]. At present, under follow-up and is disease free for more than 9 months.

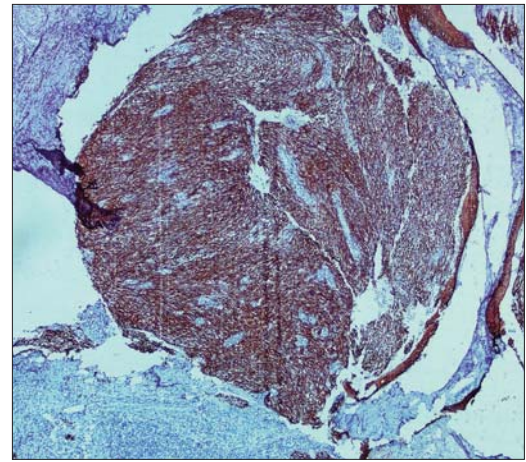


Figure 2: Immunohistochemistry staining positive for pan-cytokeratin (pan-CK) ($\times 10$)

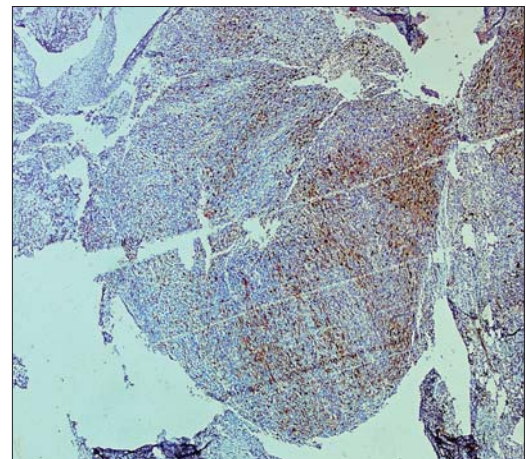


Figure 4: Immunohistochemistry staining positive for S-100 ($\times 10$)

DISCUSSION

SaC is a rare form of squamous cell carcinoma (SqCC) consisting of an epithelial component with poorly differentiated squamous and a mesenchymal component with spindle cell proliferation¹⁻⁵ with both components arising monoclonally from a single stem cell.⁵ The sarcomatoid component represents a dedifferentiation from the squamous cells and known by various names as spindle cell carcinoma, carcinosarcoma, or pseudosarcoma.^{1,5} Apart from HN, SaC has been reported in most commonly in lung followed by urinary bladder, upper aero-digestive tract, breast, gallbladder, small intestine, colon, ovary, and prostate.¹ Larynx is the most common location in HN SaC¹⁻⁵ with the most common sub-site being glottis followed by buccal mucosa in oral cavity.^{2,5} Hypopharynx as an epicenter for HN SaC has been very rarely reported and to name a few; Viswanathan *et al.*⁵ reporting six cases, Sharma



Figure 5: Whole body 18-fluorodeoxyglucose positron emission computed tomography scan showing growth right pyriform sinus (blue pointer)

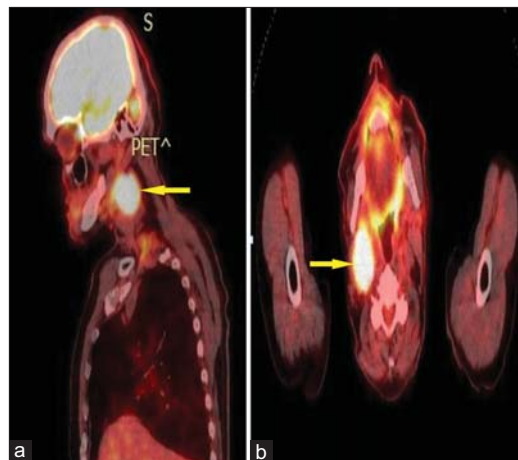


Figure 6: Conglomerate lymph node mass right neck cervical level II/III (a) sagittal section; (b) axial section (yellow pointer)



Figure 7: (a) Patient at initial presentation with swelling left side neck, (b) Patient showing significant therapeutic benefit with improvement in dysphagia and complete resolution of the lymph node mass post-chemo-radiation



Figure 8: Follow-up Whole body 18-fluorodeoxyglucose positron emission computed tomography scan 3 months post concurrent chemo-radiotherapy showing complete metabolic resolution of the growth along with cervical lymph nodes

et al.,⁷ 4 cases in Indian literature while Chang *et al.*,¹ reported 8 cases the highest till date, Iqbal *et al.*,⁴ 2 cases, Terada⁸ 1 case, Zheng *et al.*,⁹ 2 and Hardisson *et al.*,¹⁰ reported 1 case.

HN SaC has been reported mainly in elderly males^{1,2,5} between 5th and 7th decades of life,^{2,5} with risk-factors such as tobacco chewing in oral cavity,^{1,2,5} smoking and alcohol consumption in larynx¹⁻⁵ and hypopharynx,⁴⁻¹⁰ and prior RT exposure.^{1,5} Early stage lesions have been mostly reported in Western literature² as compared to advanced stages reported in developing countries⁵ due to late reporting of symptoms and lack of advanced diagnostic facilities. SaC oral cavity mostly presents as ulceroproliferative growth associated with trismus and pain,^{5,6} laryngeal lesions with hoarseness of voice while hypopharyngeal lesions cause dysphagia, odynophagia, and foreign body sensation.^{5,7-10} Lymph node (LN) involvement

is most common in oral SaC^{1,5} while least common in hypopharyngeal lesions with an incidence of 0.5%,⁵ though our case presented with N3 LN. However, as compared to HN SqCC, SaC has been found to have a higher lymphatic involvement as it can spread more easily within tissue planes which further increases the risk of distant dissemination.¹ Dissemination from hypopharynx has been to brain and skin¹⁰ while other HN sites spread to lung, liver, bone, soft tissue, skin and heart.¹

SaC has a biphasic nature comprising of the sarcomatous spindle cells which are arranged in various patterns such as fascicles as seen in our case and also storiform pattern^{2,5} and epithelial component.^{1,2,5} Tumor cells show prominent eosinophilic nucleus with high mitotic counts of 3–8/hpf with bizarre atypical mitosis.⁵ The epithelial component

may be in the form dysplasia or carcinoma-in-situ or component of the infiltrating tumor. IHC differentiates a SaC from other malignant mesenchymal tumors such as leiomyosarcoma, myoepithelial carcinoma, mucosal spindle cell melanoma, and osteosarcoma. IHC strongly positive for epithelial markers such as pan-CK and p63 and weekly positive for S-100 confirms the diagnosis of SaC while negativity for SMA, desmin, vimentin, calporin, epithelial membrane antigen (EMA) rules out other pathologies.^{2,5} Due to rarity, the debate regarding its optimal management continues. Few authors have used upfront surgery (near total hypopharyngectomy) followed by adjuvant CCRT⁹ or only RT^{1,9} while we exhibited upfront CCRT.

CONCLUSION

Attempts to understand the development, progression, and management of this tumor have been non-beneficial due to the lack of large clinical trials. Identification of unique chromosomal abnormalities, biologic pathways are necessary for the sequencing of events associated with its evolution and the development of novel biological targets for therapy. SaC should always be considered in patients presenting with a PFS lesion so as to enable a prompt initiation of appropriate treatment. We also emphasize the role of CCRT in all cases of PFS lesions with a significant clinical benefit without the morbidity of extensive surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Acknowledgment

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Conflicts of interest

There are no conflicts of interest.

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