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Sarcomatoid Carcinoma of Mid Thoracic Esophagus – A Rare Variant in Esophageal Cancers

Niaz Hussain Soomro[1], Umm-e-Habiba Abdul Rauf[1*], Mehwish Haneef[1], Qurratulain Chundriger[2] and Karim Rizwan Nathani[1]

[1]Department of Thoracic Surgery, Ojha Institute of Chest Diseases, Dow University of Health Sciences, Karachi, Pakistan.
[2]Department of Histopathology, Agha Khan University Hospital, Karachi, Pakistan.

Authors' contributions

This work was carried out in collaboration among all authors. Author NHS designed the study and wrote the protocol. Author UEHAR wrote the first draft of the manuscript and managed the literature searches. Authors MH and KRN managed the case summary. Author QC worked on the histopathology section of this report. All authors reviewed and approved the final manuscript.

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Case Report

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ABSTRACT

Aims: Sarcomatoid squamous cell carcinoma is the rarest variant of squamous cell carcinoma, which is more common at layrnx, hypopharynx, nasal cavity or oral cavity and only rarely encountered in the esophagus. Hence we present a case of sarcomatoid squamous cell carcinoma of mid esophagus which was difficult to diagnose and manage because of its rarity in our region.
Case Presentation: We report a case of sarcomatoid squamous cell carcinoma of mid esophagus which was initially diagnosed as gastrointestinal stromal tumor (GIST) of esophagus on endoscopic biopsy. After discussion with the multidisciplinary team of tumor board, McKeown esophagectomy was done. Final histopathology of the resected specimen turned out to be a sarcomatoid squamous cell carcinoma of the esophagus.
Discussion: Sarcomatoid carcinoma (SC), also known as polypoid squamous cell carcinoma or spindle cell carcinoma (SpCC), is a rare variant. It has dual histopathological appearances, i.e., either dysplastic squamous epithelium with an invasive spindle cell component, or dual epithelial

**Corresponding author: E-mail: drhabiba290@gmail.com;*

and spindle cell morphology within the invasive tumor. Data published till date mostly have described this variant of squamous cell carcinoma according to histopathological perspectives. In this case report, we have described this rare variant, which will add to the current knowledge of clinical presentation and treatment of this rare entity.

Conclusion: Limited data regarding management and prognosis of this disease, increases the necessity of further investigation on the risk factors and prognostic indicators of this disease.

Keywords: Squamous cell carcinoma; rare esophageal tumor; sarcomatoid carcinoma, spindle cell carcinoma.

1. INTRODUCTION

Esophageal carcinoma is the eighth most common cancer worldwide. It is a crippling disease with poor prognosis, as is the sixth most common cause of cancer related mortality [1-6]. It has only 15-30% 5- year cancer survival rate [7]. The most commonly seen types of esophageal cancer are squamous cell carcinoma and adenocarcinoma [7].

Sarcomatoid carcinoma is a rare variant of esophageal cancer, accounting for only 2% of all esophageal cancers [8]. It is commonly seen at larynx, hypopharynx, nasal cavity, and oral cavity in descending order of occurrence [9,10]. This type of cancer is histologically biphasic, containing both epithelial and sarcomatoid areas, the latter being composed of malignant spindle cells [7-11]. Owing to its rare occurrence at this location, data regarding the clinical presentation and management is very little. Only a handful of detailed case reports from the clinical perspective have been published in the English literature. Most of the others have discussed the pathological perspective of sarcomatoid carcinoma, as this variant of esophageal cancer poses great diagnostic difficulty under the microscope, especially in a small biopsy.

In this case report, we present a case of sarcomatoid carcinoma of mid esophagus, which is a rare variant in esophageal cancers.

2. CASE PRESENTATION

A 55-year-old male, laborer by profession with no known co-morbid, presented in thoracic surgery outpatient clinic, with complaints of difficulty in swallowing for 3 months and documented weight loss of 14 kilograms in a month. His dysphagia was progressive. He was able to take solids with the help of liquids (grade II dysphagia). There was no complain of nausea, vomiting, pain, hoarseness, shortness of breath, cough, hematemesis, jaundice, hematochezia, melena, bleeding per rectum, diarrhea, constipation, altered bowel habits or any other symptom.

Patient was naswar addict for last 45 years. Dietary history showed increased consumption of red meat and smoked food for 30 years.

Clinical examination revealed a middle-aged male with average built and height, mildly dehydrated. No other positive findings on general physical examination was observed.

Esophagogastroduodenoscopy showed an ulcerated polypoidal mass starting from 30 cm, up to 35 cm from incisors causing luminal narrowing. Multiple biopsies of the mass were taken and sent for histopathological examination.

Haematoxylin and eosin (H&E) stained slides of the endoscopic biopsy material revealed fragmented pieces of spindle cell lesion with extensive necrosis and hemorrhage (Fig. 1A). Viable areas comprised of spindle shaped neoplastic cells with abundant eosinophilic cytoplasm and markedly pleomorphic hyperchromatic nuclei with inconspicuous nucleoli (Figs. 1B and 1C). Mitotic activity was low. There was no intracellular mucin on staining with special stain periodic acid Schiff with alcian blue. Extensive panel of immunohistochemical (IHC) stains were performed including cytokeratin cocktail (CKAE1/AE3), which was negative apart from a few scattered cells (figure 1D). The tumor was positive for alpha smooth muscle actin and DOG-1 showed nuclear staining. On the basis of these findings, a possibility of gastrointestinal stromal tumor was made, which is somewhat more common than sarcomatoid carcinoma in this location.

After the biopsy report, metastatic workup was done. CT chest, abdomen & pelvis showed mid & distal esophageal neoplastic lesion with circumferential enhancing and mural thickness extending from carina up to D8 level. There was evidence of mild luminal narrowing with proximal

dilatation and presence of significant surrounding fat stranding (Fig. 2a & b). No hepatic, adrenal or bone metastases were identified.

After discussing with tumor board panel, McKeown esophagectomy was performed. During procedure a tumor of mid esophagus, measuring around 8cm in craniocaudal length, was identified (Fig. 3a & b).

Histopathology reported that the resected specimen grossly showed an exophytic mass measuring 5.5 X 3 X 0.7 cm in mid esophagus, with no involvement of gastroesophageal junction. There was no gross perforation, and the proximal margin was 2.5cm and distal margin was 8 cm away from the tumor.

H&E stained slides of the tumor revealed an epithelial neoplasm. There were areas of solid growth (Fig. 4B) alongside cribriforming with basement membrane like material (Fig. 4C) and areas of cords and ribbons of tumor cells growing in a myxoid background (Fig. 4D). After extensive sampling, only single focus of spindle cell morphology (Fig. 4A) was identified in the grossly polypoid area of tumor, which matched the morphology in the endoscopic biopsy.

Embedded within this area, there were occasional scattered nests of malignant epithelial cells with central keratinization (Fig. 5A, arrow). Cytokeratin cocktail CKAE1/AE3 was diffusely positive in all areas (Fig. 5B) and the epithelial nests within the spindle cell growth (Fig. 5C, arrow). CK5/6 and P40. IHC for DOG-1, CD117, P16, CD34, Synaptophysin and Chromogranin were all negative in the tumor. The area of spindle cell growth showed positivity for smooth muscle actin, while the epithelial nests were negative (Fig. 5D, arrow), as was seen in the endoscopic biopsy, however, DOG1 was entirely negative on this tissue. The tumor exhibited invasion in to the muscularis propria of esophageal wall along with lymphovascular and perineural invasion. The adventitial margin was 0.1 cm away. 05 recovered lymph nodes were all negative for tumor metastasis. Final diagnosis of Sarcomatoid Squamous cell carcinoma, grade III, pT2, N0 of esophagus was made.

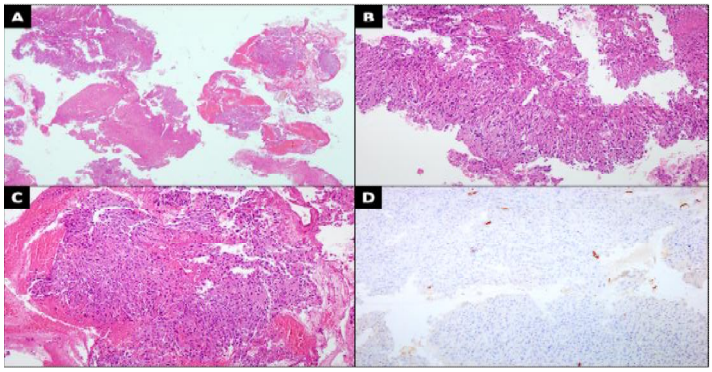

Fig. 1a, b & c: (H&E) stained slides showing spindle cell neoplastic cells with abundant eosinophilic cytoplasm and markedly pleomorphic hyperchromatic nuclei. Figure 1d: cytokeratin cocktail (CKAE1/AE3) stain negative

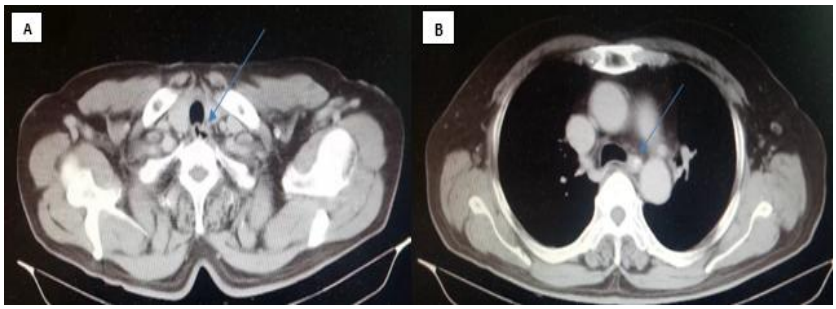

Fig. 2a. Dialated esophagus and Fig. 2b. Showing growth in esophagus
CT scan chest with IV contrast showing

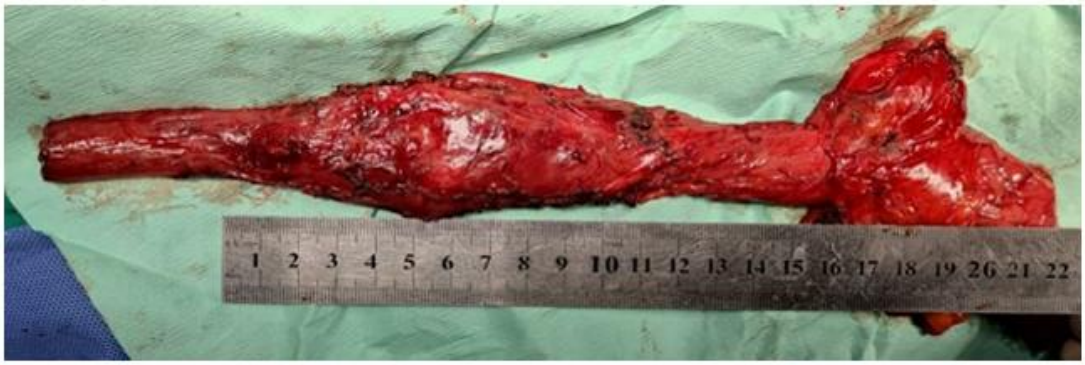

Fig. 3. Resected specimen showing mid esophageal tumor

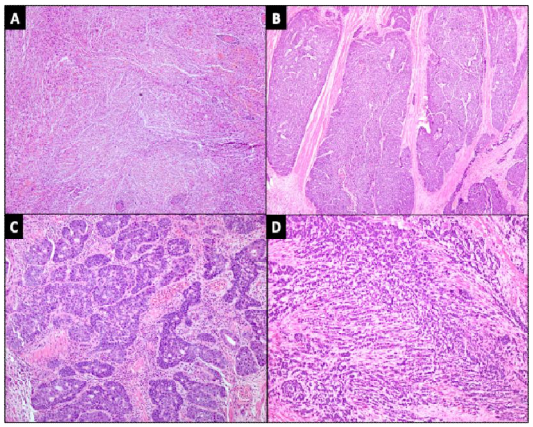

Fig. 4A: only single focus of spindle cell morphology. Fig. 5b & c: (H&E) stained slides showing epithelial neoplasm with solid growth alongside cribriforming with basement membrane. Fig. 5d: showing areas of cords and ribbons in a myxoid background.

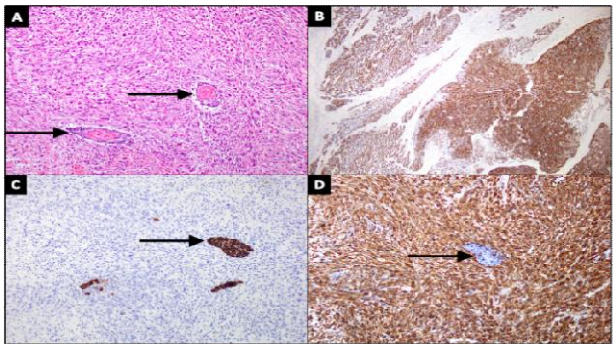

Fig. 5a. Arrow showing occasional scattered nests of malignant epithelial cells with central keratinization. Fig. 5b. showing CKAE1/AE3 stain diffusely positive. Fig. 5c. arrow showing epithelial nests within the spindle cell growth. Fig. 5d. arrow showing area of spindle cell positive for smooth muscle actin while negative in epithelial cells

Post operative recovery of the patient was smooth without any complications. After surgery patient was on jejunostomy feed. Oral feed was started on 7th post-operative day. Feeding

jejunostomy tube was removed once patient started to tolerate orally. Keeping in view the aggressive nature of this tumor, tumor board panel decided that patient must receive radiation. Hence patient was referred to the department of radiation and oncology. Patient was given radiation therapy and he tolerated the complete course. Later, patient came on 3 months follow up and was healthy from surgical aspects. Post operative wounds were completely healed. Patient was advised to follow up further with radiation oncologist.

3. DISCUSSION

Sarcomatoid carcinoma (SC), is a rare form of squamous cell carcinoma, also named as polypoid squamous cell carcinoma or spindle cell carcinoma (SpCC) [10]. Out of all esophageal malignancies it accounts for 0.5–2.8 % [11]. This rare and controversial variant has dual histopathological characteristics, i.e., dysplastic squamous epithelium accompanying with an invasive spindle cell component [7-12]. Sarcomatoid carcinoma of esophagus usually presents as an exophytic and polypoid growth. As compared to other types of esophageal cancer, it usually does not invade deeper layers of esophageal wall, thereby making it potentially resectable resulting in good prognosis [13,14].

Studies have shown that mostly esophageal carcinosarcomas occurred in old age, with male predominance. This tumor is usually prone to locoregional lymphatic metastasis with high chances of spread if not diagnosed early, with a recurrence rate of 45% [14-15].

In contrast to the above mentioned studies, Benjamin et. Al [16] and Zia et. Al [17] reported sarcomatoid SCC in elderly females. Such reports raise a possibility of a different etiological mechanism being responsible for the malignancy, rather than conventional risk factors that have been classically and extensively described in the textbooks and large studies.

Few molecular studies have evaluated the presence of certain genetic mutations such as those involving NLR and ZEB 1 genes as potential prognostic factors for esophageal sarcomatoid squamous cell carcinomas, but promising results could not be obtained due to the rarity of this disease variant [14].

Treatment modalities of sarcomatoid SCC have not been specified yet. Surgical resection is the most common approach as the tumor is mostly resectable but not in every instance. Neoadjuvant and adjuvant chemotherapy and radiation have also been considered but very less data is available regarding this. Early deaths reported are mostly due to complications associated with reconstructive surgeries [16-17]. 3-year survival is better for sarcomatoid SCC as compared to conventional type of SCC, with no significant difference in survival between the two types at 5-years [1]. Hence concluding that sarcomatoid SCC of esophagus is potentially curable. Further studies need to be done in order to establish treatment guidelines.

5. CONCLUSION

The possibility of an esophageal cancer being a sarcomatoid carcinoma should, nonetheless, be always kept in mind, particularly if the tumor appears polypoidal endoscopically and grossly.

Such rare occurrences should also be reported not only to describe the clinical and pathological findings, but also to share the follow up information, which aids in building a foundation for further large-scale studies.

CONSENT

All authors declare that ‘written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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