Case report

# Sarcomatoid carcinoma of mid thoracic esophagus – A rare variant in esophageal cancers

## 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 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.


*Key words: 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. It has only 15-30% 5- year cancer survival rate.[1] The most commonly seen types of esophageal cancer are squamous cell carcinoma and adenocarcinoma.[1]

Sarcomatoid carcinoma is a rare variant of esophageal cancer, accounting for only 2% of all esophageal cancers.[2] It is commonly seen at larynx, hypopharynx, nasal cavity, and oral cavity in descending order of occurrence.[3,4] This type of cancer is histologically biphasic, containing both epithelial and sarcomatoid areas, the latter being composed of malignant spindle cells [1-4]. 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.

No significant past medical, surgical or family history was identified. 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 (figure 1A). Viable areas comprised of spindle shaped neoplastic cells with abundant eosinophilic cytoplasm and markedly pleomorphic hyperchromatic nuclei with inconspicuous nucleoli (figures 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 (Figure 2a & b). No hepatic, adrenal or bone metastases were identified.

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

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 and distal margins of resection were clear.

H&E stained slides of the tumor revealed an epithelial neoplasm. There were areas of solid growth (figure 4B) alongside cribriforming with basement membrane like material (figure 4C) and areas of cords and ribbons of tumor cells growing in a myxoid background (figure 4D). After extensive sampling, only single focus of spindle cell morphology (figure 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 (figure 5A, arrow). Cytokeratin cocktail CKAE1/AE3 was diffusely positive in all areas (figure 5B) and the epithelial nests within the spindle cell growth (figure 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 (figure 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.

After surgery patient was on jejunostomy feed. Oral feed was started on 7$^{th}$ 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.

## 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).[4] Out of all esophageal malignancies it accounts for 0.5–2.8 %.[5] This rare and controversial variant has dual histopathological characteristics, i.e., dysplastic squamous epithelium accompanying with an invasive spindle cell component. [1-5] 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 [7,8].

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% [8-9].

In contrast to the above mentioned studies, Benjamin et. Al [10] and Zia et. Al [11] 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 [8].

Further studies concluded that although 3-year survival was better for sarcomatoid SCC as compared to conventional type of SCC, there was no significant difference in survival between the two types at 5-years [12].

Treatment modalities of sarcomatoid SCC have not been specified yet. Surgical resection is the most common approach as the tumor is resectable in most but not all instances. Neoadjuvant and adjuvant chemotherapy and radiation have also been considered. Further studies need to be done in order to establish treatment guidelines.

## 4. 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.

## 5. CONSENT

All authors declare that 'written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is attached for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

## 6. REFERENCES


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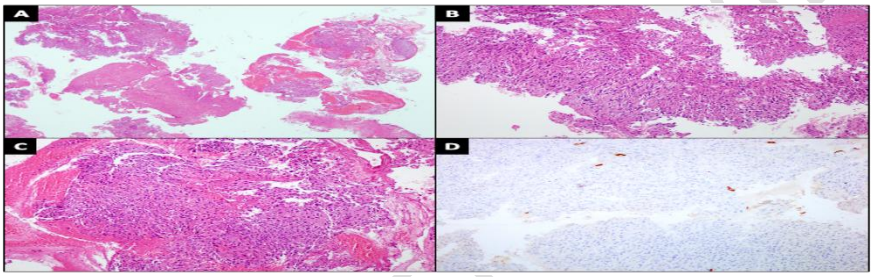
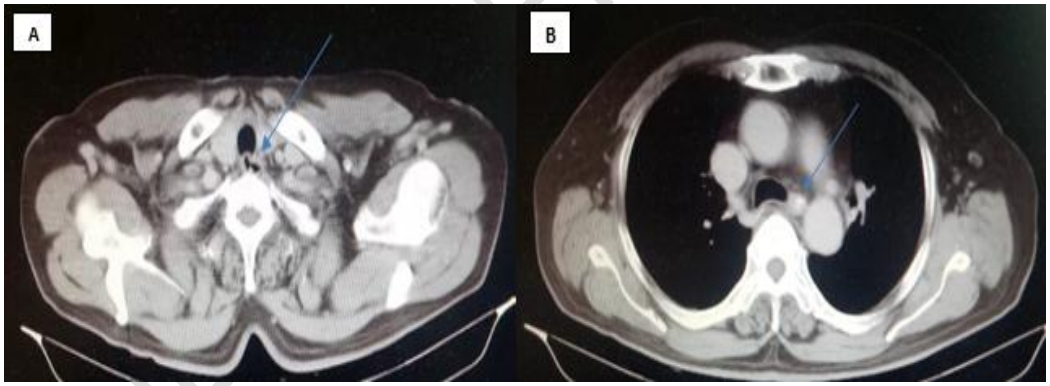
## 7. FIGURES

**Figure 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.




CT scan chest with IV contrast showing in **Fig 2a**: dialated esophagus and **Fig 2b**: showing growth in esophagus.

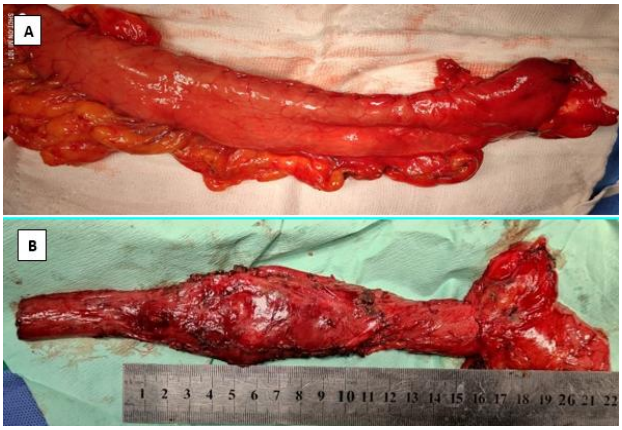



***Figure 3a**: Excised esophagus along with the tumor, where serosal layer is seen*

***Figure 3b:** Exophytic, mid esophagus tumor, when specimen was inverted, where mucosal layer is seen.*

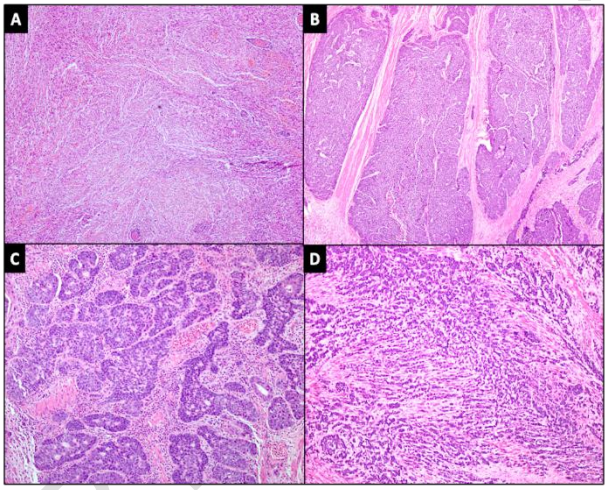



**Figure 4A**: only single focus of spindle cell morphology. **Figure 5b & c:** (H&E) stained slides showing epithelial neoplasm with solid growth alongside cribriforming with basement membrane. **Figure 5d:** showing areas of cords and ribbons in a myxoid background.

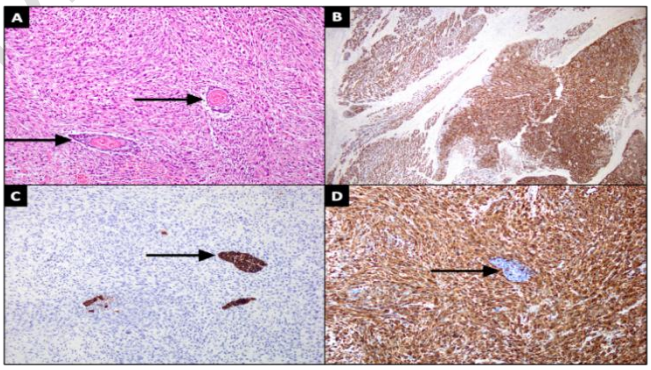



**Figure 5a:** Arrow showing occasional scattered nests of malignant epithelial cells with central keratinization. **Figure 5b:** showing CKAE1/AE3 stain diffusely positive. **Figure 5c**: arrow showing

epithelial nests within the spindle cell growth. **Figure 5d**: arrow showing area of spindle cell positive for smooth muscle actin while negative in epithelial cells.