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Carcinosarcoma of the Esophagus

Zia-ur-Rehman, Inam Pal, Zubair Ahmed* and Asim Qureshi*

ABSTRACT

Carcinosarcoma of the esophagus is a rare neoplasm characterized histologically by presence of carcinomatous and sarcomatous elements. Case report of carcinosarcoma of the esophagogastric junction whose morphological and immunohistochemical features makes it quite distinctive from other tumours is presented. It was an ulcerated lesion diagnosed in an elderly Afghan lady located 34 cms from the incisor teeth. The patient was a smoker.

Key words: *Carcinosarcoma. Sarcomatoid carcinoma. Esophagus. Smoking. Gastro esophageal junction.*

INTRODUCTION

Esophageal cancer is the sixth most common cancer in the world, especially in the Asian population.^{1,2} It is usually adenocarcinoma or squamous cell carcinoma. Composite malignant tumours with both carcinomatous and sarcomatous component are known as carcinosarcoma. A very small proportion i.e. 1.3% of esophageal tumours present as carcinosarcoma. Usually it has polypoid shape with accelerated intraluminal growth. The most consistent location described is midesophagus.³ This case of esophageal carcinosarcoma was found with ulcerative pattern at the gastroesophageal junction which was initially diagnosed as squamous cell carcinoma.

CASE REPORT

A 60-year-old lady from Afghanistan, chronic cigarette smoker, presented with progressive dysphagia for solids and weight loss for last 2 months. On examination she was malnourished with body mass index of 15.2 kg/m². There were no palpable lymph nodes. Rest of systemic examination was unremarkable. Her routine laboratory investigations were within normal range except low serum albumin of 2.7 g/dl. Upper gastrointestinal endoscopy revealed friable esophageal ulcerated mass about 34 cm from incisors teeth. Multiple biopsies of the tumour revealed it to be poorly differentiated squamous cell carcinoma. Computed tomography scan of chest and upper abdomen revealed a soft mass at gastroesophageal junction 13 mm thick (Figure 1). Cranio-caudally, it was 4 cm. The mass was separate from the aorta and pericardium. Liver was grossly normal. As the

tumour was resectable, Ivor Lewis esophagectomy was performed. Per operatively it was a well circumscribed 4 x 3 cm mass in lower esophagus at gastroesophageal junction. Histopathology of tumour revealed dual population of cells (Figure 2). Epithelial cells with squamous differentiation was showing positivity for cytokeratin AE1/AE3 on immunostaining (Figure 3). In addition, short fascicles of plump to spindle cells having moderate degree of nuclear and cytological atypia with brisk mitotic activity were also seen. This spindle cell population was positive for vimentin on immunohistochemistry (Figure 4). Features were consistent with carcinosarcoma. Four out of the 14 recovered lymph nodes showed tumour metastasis.

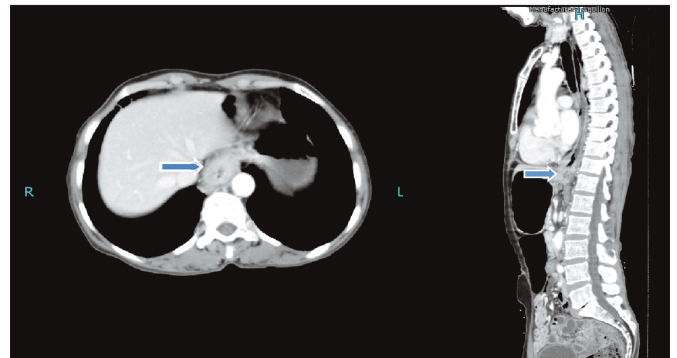


Figure 1: CT scan showing thickening at the lower end of esophagus (arrow).

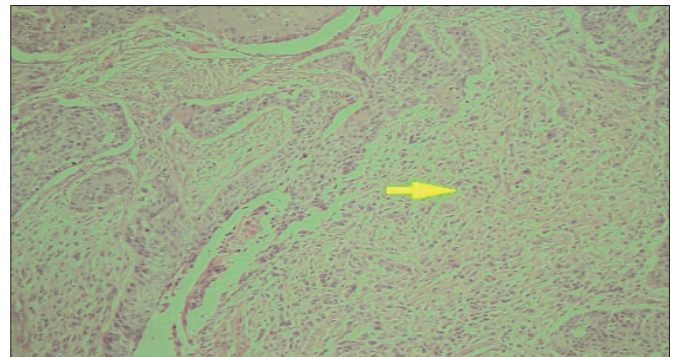


Figure 2: Histological section showing squamous cell carcinoma and sarcoma components (H and EX10).

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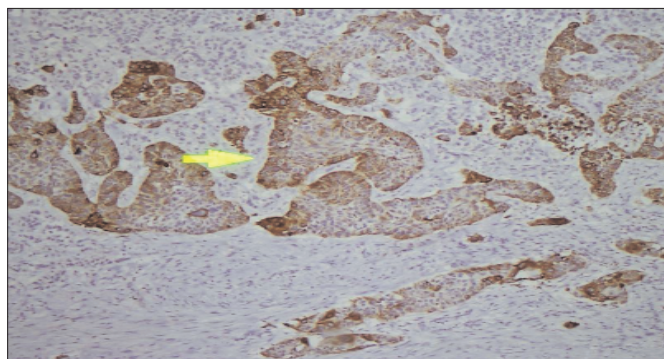


Figure 3: Positivity of carcinomatous elements for cytokeratin AE 1/ AE 3X10.

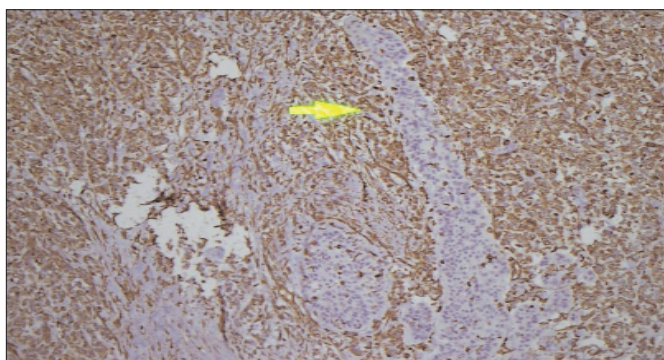


Figure 4: Positivity of sarcomatous elements for vimentin x10.

Patient tolerated the procedure well and was discharged on 8th postoperative day.

On follow-up for the last 10 months, patient was doing fine with no complains of dysphagia. There is no need of postoperative radiotherapy and chemotherapy until any evidence of recurrence of disease.

DISCUSSION

Like squamous cell carcinoma of the esophagus, carcinosarcoma occurs most often in middle-aged and elderly patients with a history of smoking or alcohol intake.

Carcinosarcoma (sarcomatoid carcinoma) usually presents as large polypoid neoplasm.^{4,5} Due to rapid intraluminal polypoidal growth, the patients present earlier.³ The most consistent location is midesophagus.³ In this case, patient was chronic cigarette smoker, but the anatomical location was different from usual.

The epithelial appearing component can be very inconspicuous and is usually limited to a few areas of *in situ* or superficially invasive carcinoma. Its appearance is usually that of a squamous cell carcinoma of either conventional or basaloid type. The bulk of the tumour has a pleomorphic sarcoma-like appearance and sometimes it exhibits focal differentiation toward cartilage, or skeletal muscle.^{5,6} In the present case, there

were demonstrations of both carcinomatous and sarcomatous component simultaneously.

According to Japanese Society for esophageal disease, three major theories have been proposed for the pathogenesis of carcinosarcoma. The first theory is that the spindle cell component is the reaction to the carcinoma.⁷ The second theory, proposes that two individual stem cells may independently and simultaneously undergo malignant transformation and are actually separate tumours that have collided (true carcinosarcoma).⁵ The third theory is that individual elements are derived from a single common ancestor cell (so-called carcinosarcoma).⁵

Ultrastructurally, some of the sarcoma-like cells in some of the tumours retain epithelial markers, such as desmosomes and tonofibrils. Most others have the appearance of myofibroblasts and other mesenchymal cells.⁸

Immunohistochemically, keratin can be consistently demonstrated in the epithelial appearing component in a high proportion of the cases and also in some sarcoma-like cells. The latter also exhibit strong reactivity for vimentin and occasionally for actin and desmin. There is a consistent overexpression of p53 as there is in conventional squamous cell carcinoma.

Because of their accelerated intraluminal growth, these tumours often present at an early stage.⁹ This may explain their more favourable outcome described in literature.

Esophageal carcinosarcoma have been treated according to the protocols used for other esophageal cancers. Treatment of esophageal carcinosarcoma does not differ from that of other malignant esophageal lesions. Surgical resection should be considered for all lesions in patients who can tolerate the operation. The indications for resectability are the same for esophageal SCC or adenocarcinoma. Additionally, the role of concomitant radiation and chemotherapy for re-irradiation of residual microscopic disease and local control must also be considered, given the unpredictable course of this tumour.

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