Epidemiology of primary small cell carcinoma of the esophagus: A retrospective study

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Dear Editor,

The common histological type of esophageal cancer is squamous cell carcinoma and adenocarcinoma. Primary small cell carcinoma of the esophagus (SCEC) is an extremely rare occurrence and not much is known about the epidemiology of SCEC from this part of the world. The age adjusted incidence rates (AAR) of esophageal cancers in both males and females is high in this part of the country and which are among the highest in the world. The AAR ranges from 27-71.4/100,000 population in males to 18.3-30.2/100,000 populations in females.[1] The prognosis of SCEC is considered to be poor. SCEC was first described in 1952 by McKeown as oat cell carcinoma of the esophagus.[2] In this analysis, we have retrospectively analyzed the gender distribution, median age, sub sites, and the median survival of patients with SCEC of our population. The data set of patients with SCEC was obtained from the cancer registry of a tertiary care cancer center in Eastern India. The data consisted of cases registered during the period from January 2010 to December 2012. The esophageal sub sites were categorized by thirds that is, upper, middle, and lower third. The follow-up for information on survival was both passive and active. The survival was estimated from the date of first diagnosis. The follow-up for survival was until March 1, 2014. The survival was calculated in months from the date of first diagnosis. Kaplan-Meir estimate was used to predict the survival probabilities of the patients and estimate the median survival period. SPSS version 19.0 (IBM Inc) was used for the present analysis.

Of 2487 (n) patients with esophageal cancers registered at our registry, a total of 11 patients with histological confirmation of SCEC were obtained from the data base. The details of gender, age and sub sites are shown in Table 1. Of 11 (n) patients, there were 8 males and 3 female patients. The age of the patients ranged from 41 to 70 years. The median age was 57 years (standard deviation = 10.5). Distribution of sub sites were in the upper third in 45% (5/11) patients, middle third in 45% (5/11) and lower third in 10% (1/11) of cases were shown in Table 1. The information on death was obtained in four patients and the rest seven patients were alive or censored at variable length of time from the date of diagnosis. The cumulative survival probability at the end of 22 months was 36% [Figure 1]. The median survival was 15 months from the date of first diagnosis.

Primary SCEC accounts for 0.5-4% of all primary esophageal malignancies. [3,4] In our retrospective analysis, SCEC accounted for 0.4% of all primary esophageal malignancies. The low proportion (0.4%) of SCEC in comparison with overall primary esophageal malignancies is significant in view of high AAR for esophageal cancers in our population. Previous reports showed male to female ratio varies from 3.2:1 to 1.5:1.[3,5] In our series of 11 patients, the male to female ratio was 2.6:1, which was in between the previous reports. The median age in a previous report was shown to be 56 years of age, [5] which was similar to the median age (57 years) in our analysis of patients with SCEC. Most of the cases of SCEC in the series of casas *et al.*

Table 1: The characteristics of 11 patients with primary SCEC

Patient	Age	Gender	Sub site	Survival (days)
1	41	Male	Middle third	26
2	45	Male	Middle third	14
3	45	Male	Upper third	289
4	70	Female	Lower third	45
5	64	Male	Upper third	27
6	55	Male	Middle third	71, dead
7	55	Male	Upper third	465, dead
8	57	Female	Middle third	386, dead
9	58	Male	Middle third	686
10	70	Female	Upper third	56, dead
11	70	Male	Upper third	608

SCEC=Small cell carcinoma of the esophagus

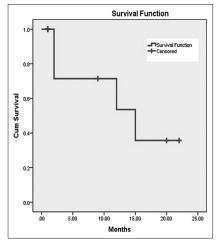


Figure 1: The Kaplan–Meir survival probability of 11 patients with small cell carcinoma of esophagus

were seen at the mid and lower third and Hudson et al. have shown that there was no case of SCEC in the upper third of esophagus. [3,6] This is in complete contrast to our series where 45% of patients with SCEC were at the upper third of esophagus and only in approximately 10% of patients had SCEC at lower third. SCEC is an aggressive malignancy with poor prognosis. The median survival in patients with SCEC depended upon the treatment received by the patient and it ranged from 7 to 18 months.^[5,7-9] However, in this series no such attempt was made to estimate the differential survival based upon the treatment received by these patients. As any prospective clinical studies of this rare entity is not feasible, hence information from large retrospective series from referral centers with correlative data are imperative.[10] In our series, the survival probability at 22 months from date of diagnosis was <36% with a median survival of 15 months from the date of first diagnosis. Furthermore, our analysis has shown that in comparison with different population groups the sub sites for the occurrence of SCEC will be different, and upper and middle thirds are common sub sites to develop SCEC in our population.

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Manigreeva Krishnatreya, Amal C. Kataki, Jagannath D. Sharma¹, Bibhuti Bhusan Borthakur², Manoj Kalita Departments of Cancer Epidemiology, ¹Pathology and ²Surgical Oncology, Dr. B Borooah Cancer Institute, Guwahati, Assam, India

> Correspondence to: Dr. Manigreeva Krishnatreya, E-mail: mani_greeva@yahoo.co.in

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