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NEUROENDOCRINE CARCINOMA OF ESOPHAGUS: A RARE CASE ENTITY

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ABSTRACT

Background: Neuroendocrine carcinoma of esophagus is a very rare and aggressive cancer. The incidence of neuroendocrine carcinoma of esophagus is 0.4% and 2% of all the esophageal malignancies. It is often diagnosed at advanced stage and has a poor prognosis.

Case Presentation: A 54-year-old male presented with a 3-month history of dysphagia and had been diagnosed with neuroendocrine carcinoma of esophagus. Patient was administered radical external beam radiation therapy 40 Gy in 20 fractions over 4 weeks with 2 courses of concomitant chemotherapy (injection carboplatin 450 mg and injection 5-fluoro-uracil 1000 mg), 3 weekly. The patient had symptomatic relief after treatment but after 2 months the patient's condition deteriorated and he ultimately died 5 months after starting treatment.

Conclusion: Neuroendocrine carcinoma of esophagus is a very aggressive cancer. More research protocols & clinical trials should be explored to improvise survival in these patients.

INTRODUCTION

Neuroendocrine carcinoma (NEC) of esophagus is a very rare and aggressive cancer. It has an incidence of 0.4% to 2% among all the esophagus cancers.[1] It has two morphological subtypes - small cell and large cell type. Small cell euroendocrine carcinoma of esophagus is more common (90% of all the cases). Most of the cases were earlier recognized as small cell carcinoma or oat cell carcinoma of the esophagus.[2]

Neuroendocrine carcinoma is positive for choromogranin A, synaptophysin and CD56 endocrine markers. A Ki67 or mitotic index of 20% or more is also necessary for diagnosing NEC. Tumors with less than 20% Ki67 positivity are diagnosed as neuroendocrine tumors.[3] Neuroendocrine carcinoma of esophagus has high incidence of lymphatic and perineural invasion.[1]

Neuroendocrine carcinoma of esophagus is often diagnosed at advanced stage and thus has a poor prognosis. The treatment of NEC of the esophagus is not well defined as only a small number of cases are reported in the literature.[4]

Neuroendocrine carcinoma of esophagus can be divided into two groups: limited disease (LD) and extended disease (ED).[5] For limited disease, the patient can undergo surgical resection (esophagectomy and extended lymph node dissection) with or without adjuvant chemotherapy.[6-7] For extended disease, chemotherapy or concomitant chemo-radiation therapy is given.[7]

Here we present a case of neuroendocrine carcinoma of esophagus and discuss the management done.

CASE PRESENTATION

A 54-year-old male presented with a 3-month history of dysphagia and had been diagnosed with carcinoma of esophagus. A previous upper gastrointestinal endoscopy had revealed a 50-mm tumor in the mid-thoracic esophagus, located 25 cm from the incisors. The pathological findings from the biopsy specimens revealed neuroendocrine carcinoma of esophagus. On immunohistochemistry, choromogranin A and CD56 tumor markers were positive. Contrast-enhanced computed tomography (CT) of chest and abdomen revealed the presence of circumferential mural thickening (25 mm thickness) involving the proximal and middle thoracic esophagus extending from 5 cm above the carina for a vertical length of 90-mm. Discrete lymph nodes were also seen in the right proximal trachea-esophageal groove. The patient had a 30-year history of smoking (30 pack years). The patient did not have a history of any chronic illness or history of cancer in the family and was not receiving any medication.

General physical examination and systemic examination were normal. Local examination of abdomen revealed scaphoid abdomen with no tenderness or organomegaly. Hematological and biochemical profile was within normal limits. Chest X-ray was also normal.

Patient was administered radical external beam radiation therapy (EBRT) 40 Gy in 20 fractions over 4 weeks with 2 courses of concomitant chemotherapy with injection carboplatin 450 mg and injection 5-fluoro-uracil 1000 mg intravenously, 3 weekly.

The patient had symptomatic relief and dysphagia was relieved after treatment initially. But after 2 months the patient's condition deteriorated and he ultimately died 5 months after starting treatment.

DISCUSSION

Neuroendocrine carcinoma of esophagus is a very rare and aggressive cancer. It has two morphological subtypes - small cell and large cell type. Small cell euroendocrine carcinoma of esophagus is more common (90% of all the cases). Neuroendocrine carcinomas are positive for choromogranin A, synaptophysin and CD56 endocrine tumor markers. Neuroendocrine carcinoma of esophagus has high incidence of lymphatic and perineural invasion.[1] Neuroendocrine carcinoma of esophagus is often diagnosed at advanced stage and thus has a poor prognosis. The treatment of NEC of the esophagus is not well defined as only a small number of cases are reported in the literature.

The present case involved a good subjective regression of disease and symptomatic relief of dysphagia after receiving radical external beam radiation therapy (EBRT) 40 Gy in 20 fractions over 4 weeks with 2 courses of concomitant chemotherapy with injection carboplatin 450 mg and

injection 5-fluoro-uracil 1000 mg intravenously, 3 weekly. But after 2 months the patient's condition deteriorated and he ultimately died 5 months after starting treatment suggesting that neuroendocrine carcinoma of esophagus is a very aggressive cancer. More research protocols & clinical trials should be explored to improvise survival in these patients.

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