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Small Cell Carcinoma of Oesophagus: A Rare Case Report and Literature Review

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Abstract **Background:** Primary small cell oesophageal carcinoma (SCOC) is a rare, highly aggressive tumor with high incidence of metastasis and associated with poor outcome. **Case report:** A 55 year old Malay lady presented to HUSM with progressive dysphagia associated with symptomatic anemia. Clinically, patient was pale and there was mass palpable at epigastric region. Biopsy revealed findings of tumor cells arranged in solid sheets exhibiting mild nuclear pleomorphism having round and hyperchromatic nuclei, inconspicuous nucleoli with frequent mitotic figures. Histopathological examination confirmed the diagnosis of SCOC. She was planned to start on CDDP/ETO for 3-4 cycles and planned for repeat CT after chemotherapy to assess the response. Unfortunately, she was only able to complete her first cycle chemotherapy partially as she had persistent low haemoglobin secondary to ongoing bleeding from tumor. **Conclusions:** The optimal treatment for SCOC is unclear. It is wise to consider radical surgery to LD patients who is fit.

Keywords Primary small cell oesophageal carcinoma, Dysphagia

1. Introduction

Primary small cell oesophageal carcinoma (SCOC) is a rare tumor which accounting for between 0.5 and 2.4% of all primary oesophageal malignancies [1]. Since it was first described in 1952 by McKeown [2], fewer than 300 cases have been reported worldwide in the literature. It is a highly aggressive, high incidence of metastasis disease at presentation which associated with poor outcome. It shares many clinical pathologic features with small cell carcinoma of lungs and other extrapulmonary small cell carcinomas are breast, ovary, uterine cervix, liver, salivary gland, stomach, colon, prostate, urinary bladder and kidney. Reported cases have been treated by various mode of treatment including surgical resection, chemotherapy, radiotherapy or combination of these methods but still no establish optimum treatment strategy has been defined. A retrospective study reporting median survival rates among 151 SCOC patients was 10.7 months where mean survival rates after 6, 12,24,36, and 60 months was 86.6%, 56.7%, 24.8%,17.4, and 12% respectively. Similarly, survival of patients receiving chemotherapy based comprehensive treatment was 12.3 months [3].

The current case study reports a case of SCOC and serves as literature review for its management and treatment.

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2. Case Report

A 55 year old Malay lady presented to HUSM with progressive dysphagia associated with symptomatic anemia. Patient reports of dysphagia, significant loss of weight and lethargy two months ago. Clinically, patient was pale and there was mass palpable at epigastric region. Hemoglobin was 5.6g% with blood pictures denotes hypochromic microcytic anaemia. Her white blood cells were normal: 6.3. She was cachexic and dehydrated with electrolytes deranged: urea 15, Na-129mmol/L, K- 3.2mmol/L, Creat 167. Apparently liver function test was normal except for hypoprotenemia.

OGDS revealed a malignant look, circumferential mass at oesophagogastric junction extending to lesser curvature. This exophytic tumour measures 4×3 cm, ulcerative with central necrotic features. There was old blood stale blood surrounding the tumour and that could be the cause of low Haemoglobin level. The tumour could have bled. Biopsy revealed findings of tumour cells arranged in solid sheets exhibiting mild nuclear pleomorphism having round and hyperchromatic nuclei, inconspicuous nucleoli with frequent mitotic figures. Immunohistochemical staining showed positive for NSE, Synaptophysin and weakly positive for Chromogranin. The histopathological examination confirmed the diagnosis of SCOC.

CT scan thorax and abdomen was performed for staging of disease which showed a tumor at cardia, lesser curvature and fundus of stomach with multiple liver and nodal metastasis involving para-aortic and subcarina lymph nodes. We resuscitated and optimized her condition, transfused 4 units of Packed cell, and albumin, and slowly rehydrated her. We started her on partial parenteral nutrition and her condition improved gradually over two weeks. Bleeding has seized and her Haemoglobin improved to 10.2g%. Renal function was back to normal.

In view of the advanced disease and unresectable tumour, patient was referred to oncology HUSM for palliative chemotherapy. She was planned to start on CDDP/ETO for 3-4 cycles and planned for repeat CT after chemotherapy to assess the response. Unfortunately, she was only able to complete her first cycle chemotherapy partially as she had persistent low haemoglobin (Hb 4.1 g%) secondary to ongoing bleeding from tumour and she was having heamatochezia. In addition, we noticed that she was leucopenic by the end of the first cycle: WBC – 2.1. She was complaining of flu like symptoms, fatigue, athralgia and nausea. All these could be attributed to the chemo drug side effects especially CDDP- cisplatin based. The medical oncologist started her on folic acid for her low Hb and olesentron for her severe nausea and vomiting. Her condition deteriorated and she passed away at home two months after the initial diagnosis of SCOC.

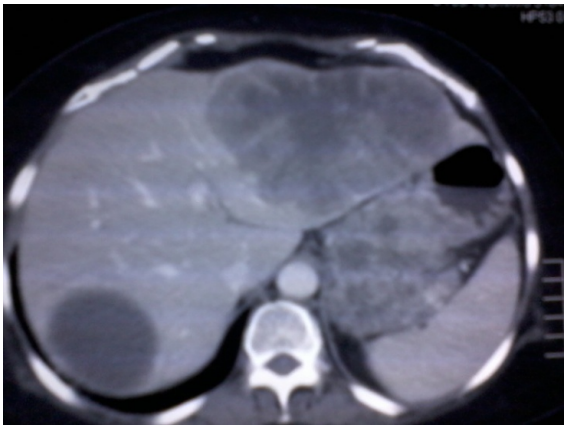


Figure 1. CT scan revealed the tumour involving the stomach with multiple liver metastases

3. Discussion

Small cell carcinoma is an aggressive tumor most frequently involved bronchial tree, which is about 15-20% of all lung cancers. It is rarely involved extrapulmonary, but reported SCOC shared the clinical behavior of small cell lung carcinoma. Microscopically, SCOC was similar to small cell lung cancer. According to 2000 WHO histological criteria for SCOC, it consisting of small, round, ovoid orspindle-shaped cells with scant cytoplasm, ill-defined cellborders, finely granular nuclear chromatin, and absent or inconspicuous nucleoli with immunohistochemical evidence of neuroendocrine differentiation.

The cellular origins of SCOC have been the subject of intense debate. It was initially thought to arise from neuroendocrine cells in submucosa which called amine precursor uptake, decarboxylation (APUD) cells. But it now appears that SCOC is endoderm in origin from the multipotent reserve cells. SCOC has potential for further differentiation into either mucin-producingor keratin-forming cells. The present study observation of dual or multiple cell types, such as the coexistence of squamous elements provides evidence of derivation from multipotent reserve cells [4]. Other studies also found the transition between small cell carcinoma and squamous cellcarcinoma in complicated SCOC [5].

The optimal treatment for SCOC is still unclear. No agreement regarding the best treatment as no large scale study done due to lower incidence rate and limited knowledge on the disease. Reviewed of few literatures show overall poor prognosis. As general rule as in other types of cancer, the staging of disease is an independent prognostic factor [6]. Most of the literatures use the staging analogous to that used in small cell lung carcinoma according to the Veterans' Administration Lung Study Group staging system (VALSG) which consists of two staging categories, limited disease (LD) and extensive disease (ED).

The largest retrospective analysis has been carried out by Casas [1] which involved the analysis of 199 patients. The literature reported a male / female ratio of 1.57:1, with 95% of tumors situated in the mid- and lower oesophagus. They reported a median survival of only 8 months for patients with LD and 3 months for patients with ED.

Radiotherapy alone shows poor survival rate as well [7]. Recent literatures suggest the use of systemic chemotherapy in the treatment of SCOC. Cases reported a median survival of 20 months in those patients receiving systemic chemotherapy compared to only 5 months in those having local treatment only [1].

A large single-institutional series involved 73 patients with SCOC who had been all treated by surgery with or without chemotherapy and radiotherapy from 1984 to 2003 were analyzed retrospectively. The incidence rate of operative complication was 11.0% (8/73). Postoperative complications included anastomotic fistula (2 patients), anastomoticstenosis (2), pulmonary infection (1), phlebitis of the lower extremities (1), gastric fistula (1), and cardiac arrhythmia (1). The 1,3 and 5-year survival rates of patients in stage I and II were75.8%, 30.3%, and 18.2%; in contrast to30.0%, 0%, and 0% of patients in stage III and IV [8]. Surgery combined with chemotherapy may be an effective approach for the treatment of stage I/II disease, but the outcome of surgery in stage III disease needs further study.

Another retrospective study involved 126 patients reports [9] single therapy with osophagectomy(S) was performed in 22 cases, radiotherapy(R) in 12 cases, and chemotherapy (C) in 7cases. 4 cases were treated with S+R; 34 cases with S+C, including 2 cases of preoperative chemotherapy; 24 cases were treated with S+C+R; and 23 cases were treated with R+C. Median survival time for LD versus ED were 14.0

months versus 7.0 months. The median survival time of 14.5 months for cases who received chemotherapy was superior to that of 5.2 months for cases who did not. The responserate to chemotherapy was high about 73.5% (25 of 34 evaluated cases). They concluded that SCOC is a systemic disease, with stage and chemotherapy as independent prognostic factors. Systemic therapy based on chemotherapy with radiotherapy is recommended.

4. Conclusions

There is no consensus regarding the treatment of SCOC. However, the role of chemotherapy is well established and should be offered to both the LD or ED patients. It is wise to consider radical surgery to LD patients who is fit, since SCOC is a disease with the major symptom of dysphagia.

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