

## SMALL CELL CARCINOMA OF THE ESOPHAGUS

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### ABSTRACT:

Primary Small Cell Carcinoma (SCC) of the esophagus is a rare but very malignant tumor characterized by early dissemination and poor prognosis if untreated. It is similar in appearance and behavior to its counterpart of the lung. Combination chemotherapy is the mainstay of treatment, though the standard treatment for esophageal SCC has not been defined yet due to the paucity of cases. The prognosis is generally dismal regardless of treatment. This patient of small cell carcinoma esophagus presented with liver and lung metastases and was treated with palliative radiotherapy and combination chemotherapy.

**KEYWORDS:** Esophageal small cell carcinoma, chemotherapy, radiotherapy, metastatic disease.

Pak J Med Sci January-March 2006 Vol. 22 No. 1 88-90

### INTRODUCTION

The most common histological type of primary esophageal neoplasm is squamous cell carcinoma; other types are rare. Among them, Small Cell Carcinoma (SCC) is an extremely rare tumor and fewer than 200 cases have been reported worldwide.<sup>1</sup> SCC of the esophagus has characteristics similar to SCC of the lung: early dissemination and a dismal prognosis.<sup>2</sup> Patients usually present with widespread metastatic disease.<sup>3</sup> Small cell carcinoma of the esophagus should be regarded as a systemic disease with a high distant failure rate.<sup>1,3</sup> Treatment strategies hence must incorporate systemic chemotherapy along with radical surgery or radiotherapy as part of a multimodality approach.<sup>4</sup>

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\* Received For Publication: April 13, 2005

Accepted: September 16, 2005

### CASE REPORT

A 55-year old male presented with the chief complaints of progressive dysphagia and pain epigastrium for one and a half years. History of loss of weight and loss of appetite was present. On general physical examination, the patient was thin built and pale. The Hb was 8.0 gm%, rest of the haemogram and biochemistry profile was unremarkable. Upper gastrointestinal endoscopy revealed an ulcerated nodular circumferential growth at 33 cms beyond which the scope was not negotiable. Contrast Enhanced Computerized Tomography CECT showed diffuse circumferential mural thickening of mid-esophagus suggestive of esophageal growth with metastatic mediastinal lymphadenopathy. Multiple lung parenchymal nodular lesions in right upper lobe of lung, along with round to oval hypodense lesions in segment V of right lobe of liver were noted on CECT thorax and abdomen. Biopsy from esophageal growth showed a tumor made up of small round cells having round nucleus, scanty cytoplasm and areas of necrosis. Guided FNA from hepatic lesions showed metastatic deposits from small cell carcinoma.

Based on these findings, a diagnosis of primary small cell carcinoma esophagus with lung and liver metastases was made. The patient was given external RT 40Gy/20F/4 weeks to

the primary site followed by Inj Cisplatin and Inj Etoposide based combination chemotherapy with palliative intent; and obtained partial symptomatic relief.

## DISCUSSION

Since the first report of small cell carcinoma of the esophagus by McKeown, most publications concerning this disease have been case reports.<sup>2</sup> Small Cell Carcinoma (SCC) of the esophagus is rare, accounting for only 0.8–2.4% of all esophageal cancers.<sup>3,5</sup> Esophageal SCC is most frequently diagnosed by the presence of dysphagia. Distant metastasis is present at the time of diagnosis in 62.5% of all esophageal SCC cases.<sup>5</sup>

Esophageal SCC might present as an ulcerating hard tumor mass on the mucosal surface of the esophagus or as a polypoid infiltrative process growing in the submucosal layer without obvious ulceration of the mucosal surface.<sup>3</sup> The mean age of patients with esophageal SCC at the time of presentation, the location of esophageal SCC and presenting symptoms of esophageal SCC are similar to those of esophageal squamous cell carcinomas.<sup>3</sup> FDG-PET may be useful in the early detection of esophageal SCC. The advantage of FDG-PET is that it can be used to diagnose the original lesion and the presence of metastases in the lymph nodes and adjacent organs.<sup>5</sup>

Esophageal SCC is of endodermal origin derived from pluripotential basal epithelial cells, which serve as the common precursor for adenocarcinoma, squamous cell carcinoma and SCC.<sup>3</sup> The small cells retain their potential for further differentiation into either mucin-producing or keratin-forming cells. This explains the coexistence of small cells, squamous and glandular elements in the same lesion. Esophageal SCC may show a variety of genetic changes, including mutation of tumor-suppressor gene and loss of heterogeneity involving the genetic loci for various tumor-suppressor genes. Recent studies have shown that Micro Satellite Instability (MSI) may be more frequent in SCC of the esophagus than in squamous-cell carcinoma of the esophagus.<sup>2,3</sup>

Since the histological and clinical characteristics of SCC of the esophagus are so similar to those of the lung, SCC of the esophagus should be treated by the same regimen used for SCC of the lung, in which chemotherapy remains the mainstay of its management.<sup>1,6-8</sup> Ohmura<sup>1</sup> treated a patient of esophageal SCC with chemotherapy alone, consisting of CisDichloro Diammine Platinum (II) (CDDP) 80 mg/m<sup>2</sup> on day1 and 70 mg/m<sup>2</sup> of etoposide (VP-16) on days 1-4. The patient had a complete response to chemotherapy, with no evidence of disease for nine months, after six courses of the regimen. Casas<sup>8</sup> used a neoadjuvant polychemotherapy regimen, followed by radiation therapy and adjuvant chemotherapy. This therapeutic combination resulted in complete response of the disease, as confirmed by computed tomography, endoscopy and biopsy. The patient is currently still alive, 6 years after the diagnosis, without any sign of relapse.

The role of surgery in patients with limited disease is controversial. In selected patients with limited stage disease and invasion limited to the submucosa, surgery with curative intent should be considered as part of multimodality treatment.<sup>6</sup> Mitani<sup>9</sup> concluded that surgical resection after obtaining a complete reduction of the primary esophageal SCC by chemotherapy may achieve longer survival when there is no evidence of metastasis on further examination. Tobari<sup>10</sup> showed that the patients with esophageal small cell carcinoma treated with surgery following chemotherapy and/or radiotherapy have been reported to survive longer than those treated with chemotherapy and/or radiotherapy.

Radiation therapy is effective, as in SCC of the lung, and is now routinely used with limited disease, but because it is only a local control therapy, radiation alone is not favorable as a first treatment.<sup>1</sup> Radiotherapy has been used concurrently with chemotherapy to enhance local control. Recurrent patterns of SCC of the esophagus are different from those of the lung: in SCC of the lung, intrathoracic lesions are common while in SCC of the esophagus, the liver or other distant organs are

common sites.<sup>3</sup>

In conclusion, with SCC of the esophagus it is almost impossible to get complete healing and local treatment alone such as operation or radiation is not favorable, so it should be treated by multi-drug chemotherapy including CDDP, with or without radiation as the first line treatment.

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