

# A Rare Tumor: Primary Small Cell Carcinoma of Esophagus: Case Report

## Nadir Bir Tümör: Özofagusun Primer Küçük Hücreli Karsinomu

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**ABSTRACT** Esophageal small cell carcinoma constitutes about 1% to 1.5% of all esophageal malignancies. The most common location of esophageal small cell carcinoma is the lower third part of esophagus. Although tumors arising in this area give early symptoms, metastatic disease is usually determined at the initial presentation and the prognosis is generally poor. Our patient was admitted to the department of surgery with general fatigue, epigastric pain, dysphagia, loss of appetite and weight loss. An esophageal small cell carcinoma at an advanced stage was detected when he was evaluated. Combined chemo-radiotherapy was proposed but he declined therapy and died two months after the diagnosis. Early diagnosis and new therapy modalities are necessary in these patients.

**Key Words:** Esophageal neoplasms; carcinoma, small cell

**ÖZET** Özofagus küçük hücreli kanseri tüm özofagus malignitelerinin yaklaşık %1-1.5'ini oluşturur. Özofagus küçük hücreli kanserinin en yaygın yerleşimi özofagus alt 1/3'üdür. Bu bölgeden köken alan tümörler erken semptom vermelerine rağmen, genellikle ilk incelemede metastatik hastalık saptanır ve prognoz kötüdür. Hastamız yorgunluk, epigastrik ağrı, disfaji, iştah ve kilo azalması şikâyetleri ile kliniğimize başvurdu. Değerlendirmeler sonucunda hastada ileri evre özofagus küçük hücreli kanseri saptandı. Hastaya kombine kemoradyoterapi önerildi, fakat hasta tedaviyi reddetti ve tanı konulduktan iki ay sonra öldü. Bu hastalık grubunda doğru yaklaşım için erken tanı ve yeni tedavi modaliteleri gereklidir.

**Anahtar Kelimeler:** Özofagus neoplazmaları; küçük hücreli kanser

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**A**lthough small cell carcinoma (SCC) is a common pulmonary neoplasm, it is rarely detected in other organs including esophagus, stomach, pancreas, small intestine, nasal cavities, paranasal sinuses, larynx, hypopharynx, salivary glands, kidney, skin, urinary bladder, prostate, breast, endometrium and uterine cervix.<sup>1,2</sup>

Esophageal carcinoma is an aggressive tumor which is difficult to cure. Its incidence in the Western world is relatively modest (United Kingdom male: 14.0, female: 9.2 per 100.000 population).<sup>3</sup> Esophageal SCC constitutes only 1 to 1.5% of all esophageal malignancies.<sup>1,2</sup> Herein we report a case of esophageal SCC with advanced disease.

## CASE REPORT

A seventy-six year-old male patient with history of heavy smoking was admitted to our surgery department with general fatigue, epigastric pain, dysphagia, loss of appetite and loss of 6 kg during the last month. His complaints started six months ago. Physical examination was normal except multiple palpable lymphadenopathies (LAP) in the cervical region. Blood tests revealed anemia (9 g/dL), and elevated liver enzymes (LDH: 413 UI/L, AST: 64 UI/L).

Esophagogastroscopy was performed in order to determine the cause of his complaints. An ulcerating and fragile mass was located between 33 cm distal to the incisors and cardia, and nodulations were detected at the cardia and the corpus of stomach (Figure 1). Multiple endoscopic biopsies obtained were reported as SCC histopathologically after light microscopic and immunohistochemical examination.

Hematoxyline and eosin stained sections of the biopsies were characterized by atypical small cells with narrow cytoplasm and dark irregular hyperchromatic nuclei located beneath the intact squamous epithelium of the esophageal mucosa. Tumor cells showed facetization to each other. Immunohistochemical analysis revealed diffuse cytoplasmic ChA, membranous CD56 and cytoplasmic

Bcl-2 staining. No positivity was observed for ACTH (Figure 2).

The next step consisted of thoraco-abdominopelvic computed tomography (CT) which demonstrated a 7 cm mass (Figure 3) that was thought to originate from esophagus, located in the cardia narrowing the lumen. Left atrium was also invaded by the tumor. Additionally, multiple perivascular, aortapulmonary, subcarinal enlarged LAP and multiple hypodense metastatic lesions less than 1 cm were detected in the liver. The CT scan failed to show any pulmonary lesions.

To eliminate the question of whether the tumor originated from the lungs, a bronchoscopy was performed. It did not reveal any neoplastic lesions arising from the lungs.

The patient was diagnosed as esophageal SCC with an advanced disease. He was consulted to the medical oncology department and combination chemoradiotherapy was proposed, but he declined therapy and discharged from the hospital with nutritional support. He died at home two months after the diagnosis.

## DISCUSSION

Primary SCC involving the esophagus is a very rare condition. The majority of cases have been re-

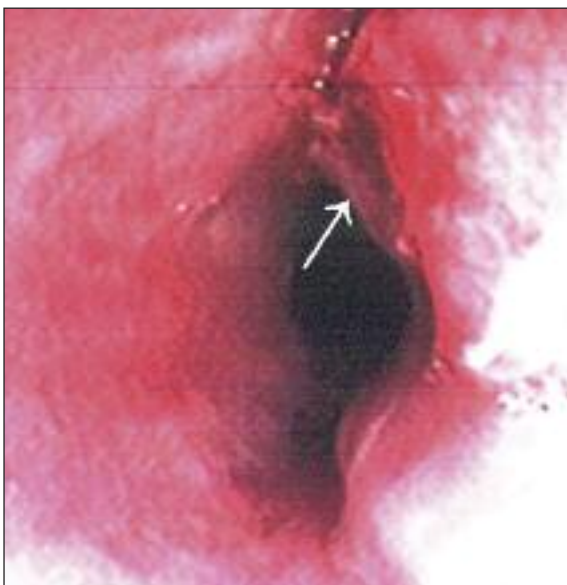


FIGURE 1: A. Ulcerating and fragile mass 33 cm distal to the incisors.

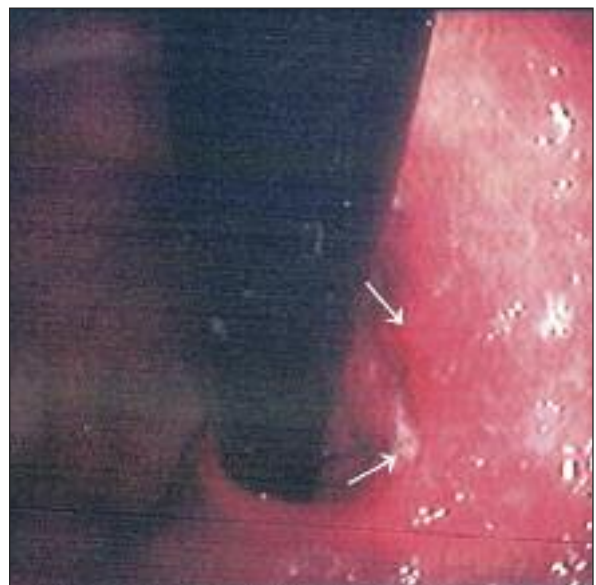
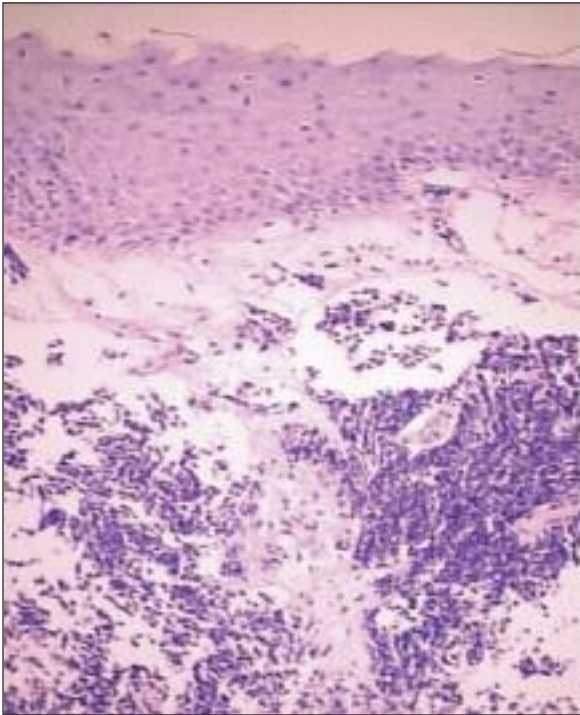
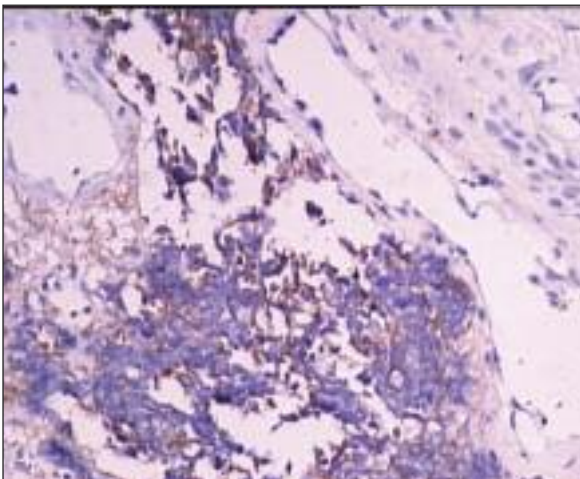


FIGURE 1: B. Nodulations at the cardia of the stomach.



**FIGURE 2: A.** Tumour infiltration beneath the squamous epithelium of the esophageal mucosa (HE, x200).



**FIGURE 2: B.** Membranous CD56 expression in tumor cells (streptavidin-biotin peroxidase, x400).

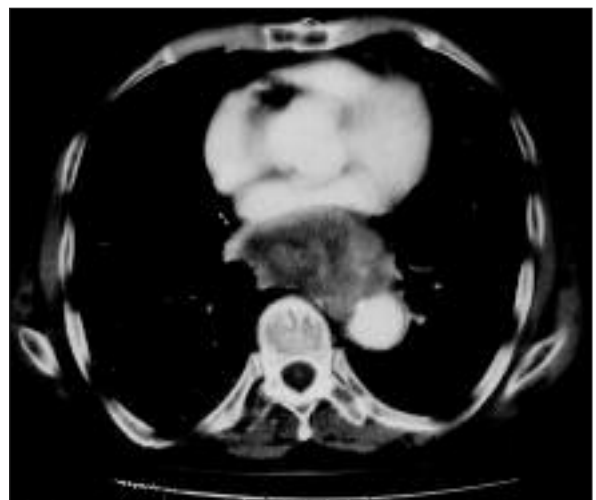
ported in males after the sixth decade and most common symptoms are progressive dysphagia, loss of appetite and weight loss. These tumors may be fungating, polypoid or ulcerative and the latter may form a fistula to the trachea. The mean age of the patients and the location of esophageal SCC are similar to those of esophageal squamous cell carcinomas. However, esophageal SCC is a more

aggressive tumor and associated with rapid growth, and patients usually present with widespread metastatic disease.<sup>4</sup>

The cellular origin of esophageal SCC is controversial. Formerly, it was thought that this tumor arises from argyrophilic Kulchitsky cells of esophageal mucosa. These cells synthesize and store amines and decarboxylate some amino acids. So, these cells are called as amine precursor uptake decarboxylation (APUD) cells. Today, it is believed that this tumor originates from pluripotential basal epithelial cells. It may present as pure SCC or as a tumor with squamous and/or glandular differentiation in microscopic analysis.<sup>2,4</sup> Esophageal SCC is an exceptional finding in patients with Barrett's esophagus, and its association with dysplastic Barrett's esophagus is extremely infrequent.<sup>5</sup>

The most common location of esophageal SCC is the lower thirds of esophagus. Although some cases may present with early symptoms, the tumors at this level usually present with extensive disease in the first evaluation. Since it disseminates early despite of different therapy modalities, the prognosis is poor.<sup>1</sup>

There is no determined advantage of one therapy modality to the others in esophageal SCC. Treatment modalities are surgery, chemotherapy, radiation therapy or any combination of these. Surgical resection was previously considered as the primary treatment of this disease. Recently so-



**FIGURE 3:** CT appearance of the 7 cm mass in the mediastinum.

me authors indicated that some patients treated with induction chemotherapy followed by chemoradiotherapy can achieve long-term survival without surgery.<sup>6</sup>

Metastases are observed usually in liver, lungs, bones and mediastinal lymph nodes. Brain metastases are uncommon in esophageal SCC.<sup>7</sup> A median survival of only eight months was reported for patients with limited disease and three months for patients with extensive disease.<sup>8,9</sup>

Our patient is a good example of esophageal SCC. He was admitted to our surgery department

with general fatigue, epigastric pain, dysphagia, loss of appetite and weight loss. These symptoms, radiologic, gastro-enterologic and pathologic work-up revealed an advanced stage of esophageal SCC. CT scan of the chest and bronchoscopy failed to show any pulmonary neoplastic lesion, and the case was therefore regarded as a primary esophageal malignancy. The prognosis of our patient was poor because he had an advanced disease. It is very hard to detect these tumors at early stages because they disseminate early. Large studies are necessary to define a treatment algorithm in these patients.

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