CASE REPORT

Small cell carcinoma esophagus: A rare entity

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Abstract

Small cell carcinoma of esophagus (SCCE), uncommon and aggressive malignancy with a poor prognosis and limited therapeutic consensus, less than 1% of all oesophageal cancers, presents with symptoms of dysphagia and weight loss, leading to diagnosis through endoscopy and histopathological evaluation.

This paper presents two case reports of SCCE. Case 1 involves a 70-year-old male with progressive dysphagia, revealing a lobulated mass in mid-esophagus diagnosed as SCCE. Imaging and biopsy confirmed the diagnosis, with positive markers for CAM5.2 and Chromogranin, managed through a multidisciplinary approach, with chemoradiotherapy initiated as primary treatment due to disease's aggressiveness and limited surgical benefit.

Case 2 describes a 60-year-old male with multiple comorbidities, demonstrated rapid disease progression and systemic complications associated with SCCE. Current literature supports the use of chemoradiotherapy for locally advanced SCCE, although overall prognosis remains poor, an average survival of approximately 12 months. Further studies are warranted to establish optimal management strategies.

Keywords: Oesophageal cancer, Oesophageal neoplasms, Small cell carcinoma, CAM5.2 Cytokeratin, Chromogranin.

DOI: https://doi.org/10.47391/JPMA.22659

Introduction

Oesophageal cancer has been ranked 8th most diagnosed carcinoma worldwide, and as 6th leading cause of death. The burden of disease is expected to rise 31.4% new cases by 2030 compared to 2020.[1] Adenocarcinoma and squamous cell carcinoma are the two most common types of oesophageal malignancy, while less than 1% of cases consist of sarcomas, lymphomas, and other rare lesions.[2] A study conducted in Pakistan found that small cell carcinoma of oesophageal origin (SCCE) is the least common type, with adenocarcinoma being the most prevalent.[3] Small cell carcinoma of oesophagus is a rare entity, aggressive and has a poor prognosis. Management of this disease is still under debate and no consensus has been made.[4] Verbal informed consent was obtained from the patients for the publication of these case reports, ensuring that no identifying information would be disclosed.

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Submission complete: 20-12-2024 **First Revision received:** 16-05-2025

Acceptance: 15-04-2026 **Last Revision received:** 14-04-2026

Case 1

A 70-year-old male, with no co-morbids and performance status of 0-1, reported to outpatient clinic of gastroenterology department in Shifa International Hospital Islamabad on 26th September, 2023 with complaints of dysphagia and chest discomfort which started two months ago. The patient reported taking small bites of food and required sips of water between bites to facilitate swallowing. There was associated odynophagia, undocumented weight loss and loss of appetite. There was no history of vomiting, haematemesis or altered bowel habits. On examination patient was lying comfortably, and had no pallor or jaundice, abdominal and lymph node examination was unremarkable, and Body Mass Index was 31.22kg/m2. He was booked for Upper Gastrointestinal endoscopy and initial blood work was done including haemoglobin which was 12g/dl. His endoscopy showed a large lobulated, friable semi circumferential mass causing some luminal obstruction in mid oesophagus at 28cm from incisors, endoscope however was negotiated through it and Gastrooesophageal Junction was at 40cm. Moderate mucosal erythema and oedema was noted in fundus and body of stomach. Rest of the examination was normal (Figure 1 A, B).

Multiple biopsies of the mass were taken and dietary advise was given. Computed Tomography scan Chest, Abdomen and pelvis was advised which showed a soft tissue density minimally enhancing lobulated mass in posterior mediastinum, in mid oesophagus, inseparable

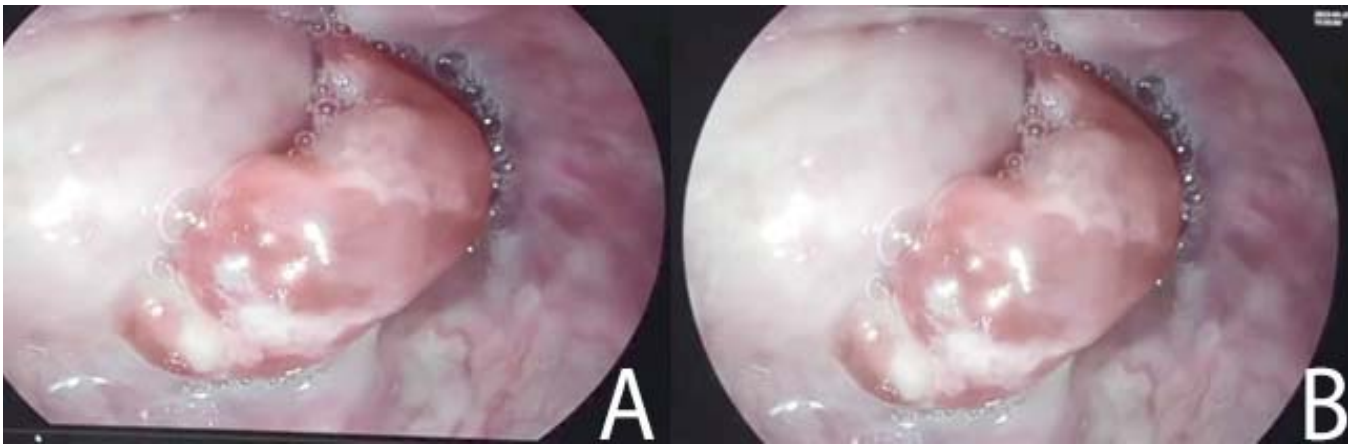

Figure-1: A, B: Large Lobulated mass.

from posterior wall causing severe luminal narrowing measuring 4.4x3x6.5cm in size. Lesion was covering 90 degrees of aorta antro-medially. There was mild pooling of contrast above the lesion in oesophagus however distal passage of oral contrast was seen in stomach. There was mild mass effect of this mass on carina and proximal left main bronchus, but otherwise the airway was patent. Multiple mildly enlarged mediastinal lymph nodes were seen with the largest measuring 11mm (Figure 2 A, B).

The histopathology report of biopsies taken from the mass showed oesophageal tumour composed of oval to spindle cells showing scant cytoplasm and moulding, suggesting small cell carcinoma. Immunohistochemistry was positive for CAM 5.2 (Cytokeratin Antibody Marker 5.2) and Chromogranin. Ki67 index was high and Synaptophysin was weak positive. P40 was negative. (Figure 3 A, B, C)

The patient was referred to Surgical Clinic, where he was evaluated and was referred to an Oncologist. His case was discussed in multidisciplinary team, and it was decided due to the locally advanced and aggressive nature of disease, surgery alone would not be beneficial. Chemoradiation followed by re-evaluation was advised.

Case 2

A 60-year-old male with a history of hypertension and hepatitis B virus–related decompensated chronic liver disease and hepatocellular carcinoma presented to Shifa International Hospital, Islamabad, on 12th July with complaints of dysphagia, nausea, and vomiting. Imaging revealed a necrotic hepatic mass in segment IV-B, extensive superior mesenteric vein thrombosis, and enlarged lymph nodes. Endoscopy of the oesophagus demonstrated a distal oesophageal mass, which was confirmed as small cell carcinoma on biopsy.

One week later, the patient developed altered sensorium and was admitted to the critical care unit with septic shock, acute kidney injury, hepatic encephalopathy, and acute-on-chronic liver failure. Laboratory investigations

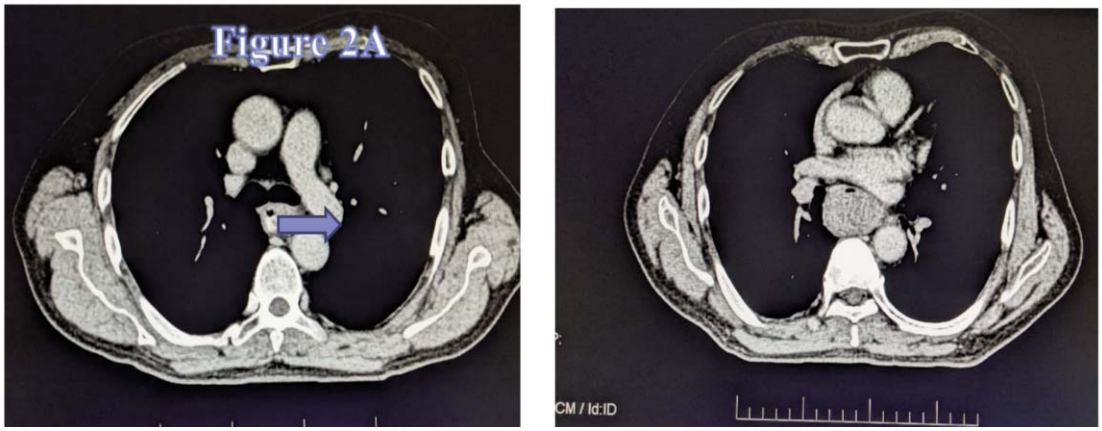

Figure-2: A, B: Mid-Oesophageal Lobulated Mass, Causing severe luminal narrowing, measuring 4.4x3x6.5cm, adjacent to the posterior wall, covering 90 degrees of the aorta antro-medially

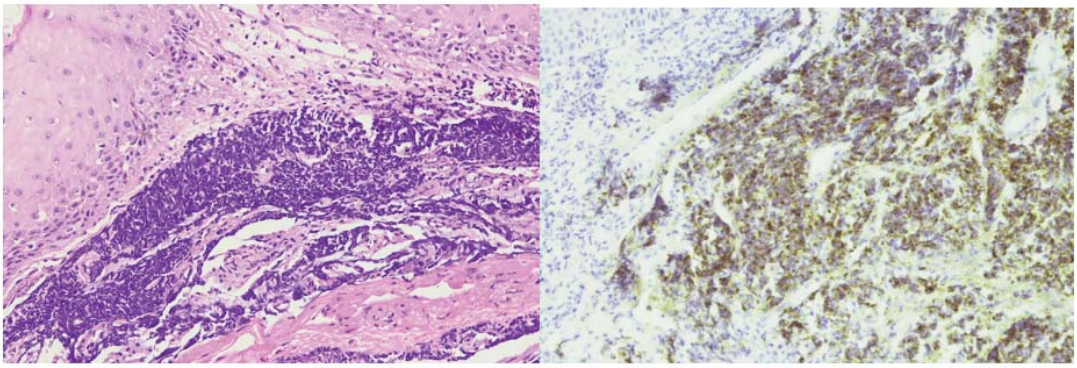

Figure 3A

Figure 3B

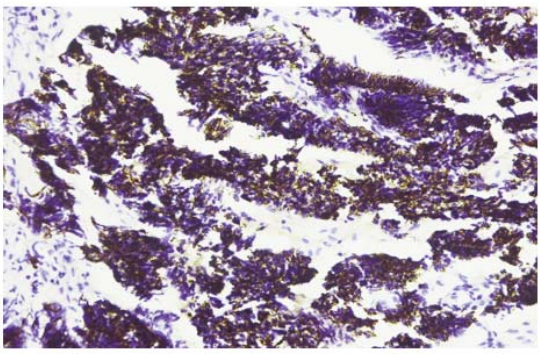

Figure 3C

Figure-3: A, B, C: Ooesophageal Mucosa with Underlying Tumour, composed of oval to spindle cells with scant cytoplasm, indicating small cell carcinoma. Immunohistochemistry Findings: Positive for CAM5.2 and Chromogranin, with high Ki67 index; Synaptophysin weakly positive, P40 negative.

showed elevated serum ammonia (ammonia= 113 µmol/L; normal = 19–48 µmol/L) and C-reactive protein levels, (CRP = 80mg/L; normal = upto 5.0 mg/L, along with hypoalbuminaemia (S.Albumin = 2.5g/dL; normal=3.5 to 5.5 g/dL). Despite aggressive management with inotropic support, haemodialysis, and broad-spectrum antibiotics, the patient's condition deteriorated, and he subsequently died.

Discussion

Small cell carcinoma is common malignancy in lung, and accounts for 14% of lung cancers.[5] Incidence of SCCE is reported to be <1% and it has been found to be an aggressive tumour with poor prognosis.[6] Most common presentation is with dysphagia and weight loss due to which patient is subjected to endoscopy and biopsy of the lesion leading to diagnosis of the disease. There are two types reported, one being small cell carcinoma of the lung and the other is ulcerative and diffuse type. No significance difference in terms of prognosis has been associated with this. For staging purposes, small cell carcinoma oesophagus is divided in two groups Local and Regional disease, with former being without distant metastasis and surrounding invasion of organs or tissue, and later with nodal involvement and or distant metastasis. Histopathology is the only modality to diagnose small cell carcinoma whereas immunohistochemistry markers Chromogranin A and Synaptophysin can be positive.[4] Due to a low incidence, there have been limited studies on the management of SCCE and no consensus has been made.[7] A retrospective analysis done by Chen et al in patients with small cell carcinoma esophagus treated with radiotherapy as part of treatment also found overall survival to be 12 months.[8]

Conclusion

Small cell carcinoma esophagus is a rare entity with an aggressive course. Management should be planned via a multidisciplinary team approach; surgical management should not be the first option and the patient should receive chemoradiotherapy. Surgical management should be reserved for patients with limited disease, as no significant difference in overall survival with different treatment modality has been observed. Further studies

are still required for management of this disease.

Acknowledgment: We acknowledge the invaluable contributions of Dr. Muhammad Bilal (Associate Consultant Histopathology) and the Department of Pathology and Laboratory Medicine at Shifa International Hospital, Islamabad, for their expert analysis and support in this case.

Disclaimer: None.

Conflict of Interest: None.

Source of Funding: None.

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AUTHORS' CONTRIBUTIONS:
SA: Concept and design.
MAA: Data collection and final approval.
FA: Data collection.
HK: Concept and supervision.
MA: Review the report.
HG: Supervision.