

## A Rare Case of Esophageal Adenocarcinoma with Signet Ring Cell Features

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## Introduction

Adenocarcinomas with signet ring cell features (SRC) features are rare gastrointestinal cancers found in the stomach and colon. Esophageal adenocarcinoma (EA) with signet ring cell (SRC) features is an extremely rare entity with a grim prognosis. We present a rare case of EA with SRC features in a patient who presented with food impaction.

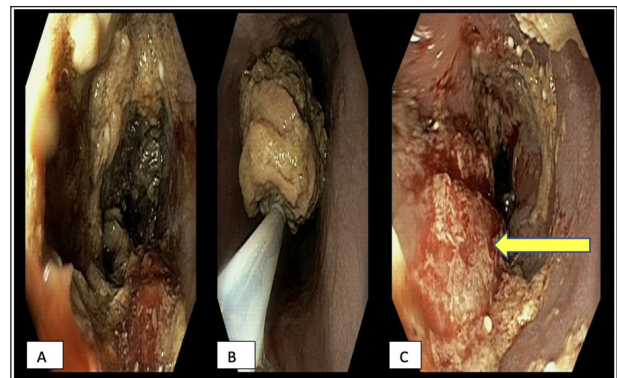
## Case Description

A 65-year old male with past medical history of hypertension presented with difficulty swallowing and regurgitation after having dinner at a steak house. Patient reported a 40-pack year smoking history and occasional alcohol use. In the ED, patient was found to be hemodynamically stable, labs unremarkable for any abnormalities. Patient was unable to swallow liquids or solids. GI was consulted for further evaluation. Patient underwent emergent EGD which showed food impaction with pieces of steak in the distal esophagus (Figure 1 A & B). Multiple pieces of steak were removed with a Roth net retriever. After removal of the impacted food, a friable, ulcerated and fungating mass was visualized in the distal esophagus (Figure 1 C) that nearly occluded the lumen. Cold forcep biopsies were taken from the mass. Given concern for malignant obstruction, an esophageal stent was placed and secured with stentfix OTSC clips (Figure 2 A & B). Patient was able to tolerate a diet post-procedure and was subsequently discharged. Biopsy results revealed moderate to poorly differentiated adenocarcinoma with focal signet ring cells (Figure 2 C). Patient was referred to thoracic surgery and oncology.

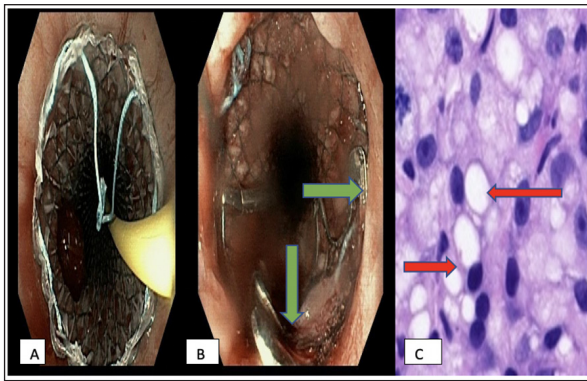
## Discussion

A signet ring cell is characterized by abundant intracytoplasmic mucin that displaces the nucleus to the periphery thereby making it resemble a "signet ring". Signature SRCs comprise majority of adenocarcinomas with SRC features. These carcinomas have

a predilection to affect the gastrointestinal tract but EA with SRC features represents a rarest of rare histologic subtype of EAs (incidence of less than 0.5% of all adenocarcinomas with SRC features and approximately 2 % of all esophageal cancers) therefore making its diagnosis elusive. Patients present with varying symptoms such as obstruction (causing food impaction as in our patient), dysphagia, odynophagia and hematemesis. There is a paucity of information about the natural history of the disease, epidemiology and risk factors. However, existing retrospective studies show that EA with SRC features portend a poor overall survival (owing primarily to advanced disease at the time of diagnosis and bleak response to treatment with surgery or chemotherapy) compared with EA with a non-SRC histology. Our case brings to light, the importance of early detection of this unique histologic variant of EA. Clinicians must assimilate the knowledge of this rare disease state in order to initiate prompt treatment with surgical resection and/or chemotherapy.



**Figure 1A:** Impacted steak seen in the distal esophagus. **B:** Impacted steak being removed from distal esophagus. **C:** Fungating and friable esophageal mass seen in the distal esophagus (yellow arrow).



**Figure 2A:** Esophageal stent being placed in the distal esophagus. B: Esophageal stent secured in place with stentfix OTSC clips (green arrows). C: Signet ring cells seen on esophageal biopsy