

# Pyopericardium secondary to achalasia-associated squamous cell carcinoma of the oesophagus

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## ABSTRACT

Patients with achalasia of the oesophagus are known to be at increased risk of oesophageal squamous cell carcinoma. To our knowledge, this is the first report of an achalasia-associated oesophageal squamous cell carcinoma presenting with acute sepsis secondary to pyopericardium.

## KEYWORDS

Achalasia – Oesophageal squamous cell carcinoma – Pyopericardium – Malignant pericardial effusion

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## Case history

A 51-year-old man diagnosed with oesophageal achalasia in 2008 presented to the emergency department with severe sepsis and multi-organ failure. He had previously declined treatment and now presented with a 6-month history of progressive dysphagia and weight loss but no other comorbidities. Computed tomography (CT) demonstrated a grossly distended mega-oesophagus (82.2mm maximum diameter) containing large amounts of food debris and a pericardial effusion (Figs 1 and 2).

In view of an evolving cardiac tamponade, he underwent urgent drainage of the pericardial effusion. Subsequent therapeutic rigid oesophagoscopy and diagnostic oesophagogastroduodenoscopy (OGD) revealed a grossly distended oesophagus, with a circumferential mass in the lower third (30–45cm), arising from the gastro-oesophageal junction (GOJ). Biopsy histology demonstrated poorly differentiated squamous cell carcinoma (SCC). Microbiology results of the pericardial fluid confirmed an infective pyopericardium with growth of Gram-negative bacilli (*Lactobacillus*) and *Candida* species. The presence of gastrointestinal flora within the pericardial sac suggests translocation of microorganisms from the oesophageal lumen into the pericardium. An alternative mechanism may involve perforation of his tumour into the pericardium, resulting in malignant pyopericardium.

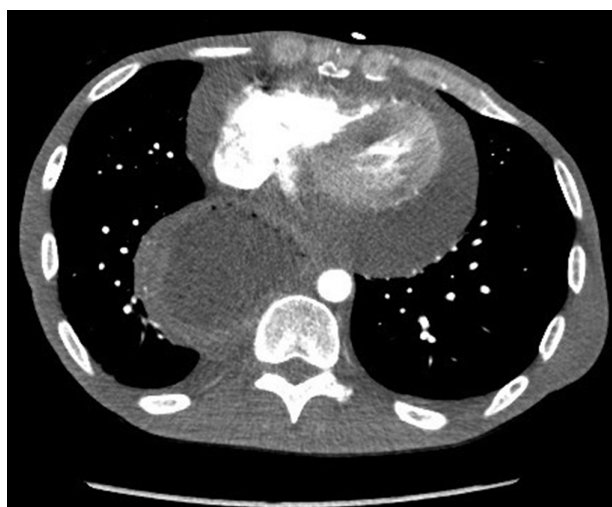
Subsequent staging CT confirmed a 3.3cm GOJ lesion with peritoneal disease and a 2.5cm left adrenal metastasis (T4N2M1). Following discussion with the multidisciplinary team and the patient, a palliative care plan was initiated.

## Discussion

Achalasia of the oesophagus is a primary motility disorder characterised by aperistalsis and incomplete relaxation of the lower oesophageal sphincter on swallowing. It has an incidence of 1/100,000 per year.<sup>1</sup> Patients with achalasia are up to 16 times more likely to develop oesophageal SCC compared with the general population. Likely aetiological factors include stagnation and chronic irritation from food debris within the oesophageal lumen.<sup>1–2</sup>

Diagnosis of achalasia-associated SCC of the oesophagus is often delayed. This is possibly because patients are already accustomed to dysphagia, meaning that affected individuals may fail to recognise progression of their symptoms over time alongside non-specific constitutional decline. Unsurprisingly, therefore, prognosis of oesophageal SCC within this patient cohort is poor.<sup>3</sup> However, liberal use of surveillance in high-risk and symptomatic patients has shown that cancers can be detected early within this patient cohort, with prognostic outcomes similar to those without achalasia.<sup>4</sup> There are currently no clinical guidelines to recommend when to refer patients with achalasia for further investigation, meaning that clinicians should operate with a high degree of suspicion when patients describe a change in symptoms.

Malignant pericardial effusions are well described in the medical literature, occurring most commonly in lung and breast cancer, and often have a metastatic or paraneoplastic aetiology. Therefore, malignant pericardial effusions are often sterile with high protein (exudate) content.<sup>5</sup> To our knowledge, this is the first case of a patient with an



**Figure 1** Computed tomography: transverse view demonstrating dilated oesophagus with large pericardial effusion



**Figure 2** Computed tomography: coronal view demonstrating grossly distended oesophagus full of food debris

underlying oesophageal SCC presenting with acute sepsis and cardiac tamponade secondary to a malignant pyopericardium.

It can be hypothesised that this unusual presentation was the result of progressive untreated achalasia with coexisting malignancy, resulting in either translocation of bacteria or perforation into the pericardial sac. While malignant perforation secondary to gradual erosion of the tumour into the pericardium is plausible, the absence of air within the mediastinum and pericardial sac on the CT makes this hypothesis difficult to prove. It is thus more likely that the dilated oesophagus and high luminal pressures allowed translocation of bacteria and subsequent pyopericardium. The usual management of patients presenting with malignant pericardial effusions following pericardiocentesis is pericardial window via either a subxyphoid or transthoracic approach.<sup>5</sup> Definitive management was not feasible in this case owing to the patient presenting in extremis with multi-organ failure.

## References

1. Sandler RS, Nyren O, Ekblom A *et al.* The risk of esophageal cancer in patients with achalasia: a population-based study. *JAMA* 1995; **274**(17): 1,359–1,362.
2. Streitz JM, Ellis FH, Gibb SP *et al.* Achalasia and squamous cell carcinoma of the esophagus: analysis of 241 patients. *Ann Thorac Surg* 1995; **59**(6): 1,604–1,609.
3. Ribeiro U, Posner MC, Safatle-Ribeiro AV *et al.* Risk factors for squamous cell carcinoma of the oesophagus. *Br J Surg* 1996; **83**(9): 1,174–1,185.
4. Br ucher BL, Stein HJ, Bartels H *et al.* Achalasia and esophageal cancer: incidence, prevalent and prognosis. *World J Surg* 2001; **25**(6): 745–749.
5. Liberman M, Labos C, Sampalis JS *et al.* Ten-year surgical experience with nontraumatic pericardial effusions a comparison between the subxyphoid and transthoracic approaches to pericardial window. *Arch Surg* 2005; **140**(2): 191–195