CHIRURGIE THORACIQUE / THORACIC SURGERY

MALIGNANT TRANSFORMATION IN ACHALASIA OF THE CARDIA AFTER TRANSTHORACIC OESOPHAGECTOMY: A CASE REPORT

MN. TAMATEY[1,3], MM. TETTEY[1,2], I. OKYERE[4], IK. ADZAMLI[1], K. ADOMAKO[1], G. OFFEI-LARBI[1]

1. National Cardiothoracic Centre, Korle-Bu Teaching Hospital, Accra, Ghana
2. Department of Surgery, University of Ghana Medical School, Accra, Ghana
3. Department of Surgery, School of Medicine, University of Health and Allied Studies, Ho, Ghana [4]Department of Surgery, School of Medicine and Dentistry, College of Health Sciences, Kwame Nkrumah University of Science and Technology, Kumasi, Ghana

Correspondence: Martin Tamatey
Email: mtamatey@yahoo.com

Abstract
Achalasia of the cardia is a pre-malignant condition. Though it is uncommon, it is known that malignant change can occur several years after the treatment for achalasia. This is because the chronic food stasis in the oesophagus over the years affects mainly the distal and mid-oesophagus. This leads to chronic irritation of the mucosa, which finally leads to the malignant change. We present the case of malignant transformation in achalasia of the cardia. A 56-year old lady presented at the National Cardiothoracic Center in December 2012 with a 2-month history of difficulty in breathing and weight loss. She had modified Heller’s operation for achalasia of the cardia 25 years earlier, and transthoracic oesophagectomy 15 years earlier, for recurrence of the achalasia, complicated by a sigmoid oesophagus. The CT scan showed a tumour in the remnant (intrathoracic) oesophagus infiltrating the trachea. Oesophagoscopy confirmed the tumour, and the biopsy came out as squamous cell carcinoma of the oesophagus. She subsequently developed dysphagia, and passed away shortly after that. The clinical significance of this case is that since the food stasis does not usually affect the cervical oesophagus, it is often spared from the malignant change. Therefore, when oesophagectomy is considered for recurrent achalasia or failed Heller's operation, it will be a better option to remove the whole of the thoracic oesophagus, and the anastomosis is done at the level of the cervical oesophagus (with the colon or the stomach). This will significantly reduce the chances of developing a malignancy, thereby sparing the patient’s life, unlike in this case. Long-term surveillance of achalasia patients is also strongly recommended.
Keywords: Achalasia, malignant transformation

Introduction

Achalasia of the cardia is a motility disorder of the oesophagus of unknown aetiology. It is characterized by failure of relaxation of the lower oesophageal sphincter during swallowing, hypertrophy and dilatation of the oesophagus, and the absence of peristalsis in the oesophagus. These factors lead to chronic food stasis in the oesophagus. The chronic food stasis, in the long term, can lead to malignant transformation. This is what makes achalasia of the cardia a pre-malignant condition, with the malignancy occurring several years after the initial diagnosis of the achalasia. The incidence of developing oesophageal carcinoma in achalasia is 3.3%.[1]

We present the case of a patient who was managed in the National Cardiothoracic Centre, Korle-Bu Teaching Hospital, and developed a malignancy 25 years after the initial treatment for the achalasia.

Case report

A 56-year old lady presented at the National Cardiothoracic Center in December 2012 with a 2-month history of difficulty in breathing and weight loss. She had had modified Heller's operation for achalasia of the cardia 25 years earlier, and transthoracic oesophagectomy 15 years earlier, for recurrence of the achalasia. It was also complicated by a sigmoid oesophagus. The CT scan showed a tumour in the remnant (intrathoracic) oesophagus, infiltrating the trachea and causing the difficulty in breathing. Oesophagoscopy confirmed the tumour, and the biopsy came out as squamous cell carcinoma of the oesophagus. She then developed dysphagia, for which a feeding jeunostomy was done. She passed away two months later.

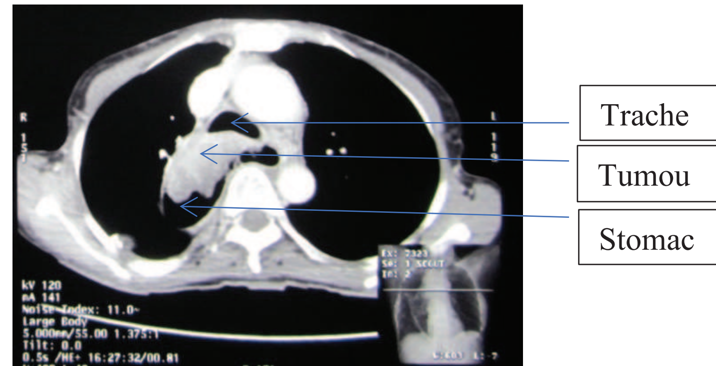

Fig. 1. *CT scan of the chest showing the tomour in the stomach and, indenting the trachea*

Discussion

Achalasia of the cardia has long been known as a pre-malignant condition, with the oesophageal carcinoma developing several years after the onset of symptoms of the achalasia. Durations varying from 5 to 24 years have been reported.[1,2,3,4] Although the prevalence of patients with achalasia developing an esophageal carcinoma is low the risk is nearly 140-fold; there is no difference in prognosis between patients with achalasia-carcinoma and those with esophageal cancer without achalasia[5]. Our patient developed the oesophageal carcinoma 25 years after the initial presentation of the achalasia. The chronic food stasis over the years affects the distal and mid-oesophagus. This leads to irritation of the mucosa, which then leads to metaplasia, dysplasia and finally the carcinoma of the oesophagus. Since the food stasis does not usually affect the cervical

oesophagus, it is often spared from the malignant change. At the second surgery, the patient had transthoracic oesophagectomy, leaving behind part of the thoracic oesophagus. This is the part that developed the carcinoma several years later. If the oesophagectomy had involved the whole of the abdominal and thoracic oesophagus, leaving behind only the cervical oesophagus, it is unlikely that a malignancy would have developed later, since the cervical oesophagus is often not involved with the chronic food stasis and irritation. It is therefore our opinion that when oesophagectomy is considered for recurrent achalasia or failed modified Heller's operation, the whole of the thoracic oesophagus must be removed, and the anastomosis done at the level of the cervical oesophagus. The oesophageal replacement can be done with the colon or the stomach. There must also be long-term surveillance of achalasia patients.

Conclusion

Malignant transformation in the oesophagus occurs several years after the initial diagnosis of achalasia. When oesophagectomy is considered in the course of the management of the achalasia, the whole of the thoracic oesophagus must be removed and the anastomosis done at the level of the cervical oesophagus, to prevent the development of malignancy in the remnant intrathoracic oesophagus.

References

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