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Malignant Transformation in the Esophagus 25 Years after Esophagectomy for Achalasia: Case Report

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ABSTRACT

Achalasia of the cardia though uncommon, can be a premalignant lesion. It is known that malignant changes can occur several years after its treatment. This is because the chronic food stasis in the esophagus over the years leads to irritation of the mucosa, which finally leads to the malignant change. A 31-year-old lady had modified Heller's operation a few months after the onset of the symptoms of achalasia. Ten years later, she presented with recurrence of the achalasia, complicated by a sigmoid esophagus. She then had transthoracic esophagectomy. Fifteen years after the esophagectomy, she presented with severe weight loss and difficulty in breathing. The CT scan showed a tumor in the remnant (intrathoracic) esophagus infiltrating the trachea. There was no dysphagia. Esophagoscopy confirmed the tumor, and the biopsy came out as squamous cell carcinoma of the esophagus. She passed away within a few days before radiotherapy could be commenced. Since the food stasis does not usually affect the cervical esophagus, it is often spared from the malignant change. Therefore, when esophagectomy is considered for recurrent achalasia or failed Heller's operation, we recommend that the whole of the thoracic esophagus be removed, and the anastomosis done at the level of the cervical esophagus (with the colon or the stomach). 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 esophagus of unknown etiology. It is characterized by failure of relaxation of the lower esophageal sphincter during swallowing, hypertrophy and dilatation of the esophagus, and the absence of peristalsis in the esophagus. These factors lead to chronic food stasis in the esophagus [1-6]. The chronic food stasis, in the long term, can lead to malignancy. 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 esophageal carcinoma in achalasia (3.3%) has been reported [1] We present the case of a patient who developed a malignancy 25 years after the initial treatment for the achalasia.

CASE REPORT

A 31-year-old lady had symptoms of achalasia. A couple of months later she had modified Heller's operation. Ten years later, she presented with recurrence of the achalasia, complicated by a sigmoid esophagus. She then had esophagectomy with esophagogastrostomy (Ivor-Lewis). Fifteen years after the esophagectomy, she presented with severe weight loss and difficulty in breathing. The chest CT scan **Figure 1** showed a tumor in the remnant (intrathoracic) esophagus infiltrating the trachea. There was no dysphagia. esophagoscopy confirmed the tumor, and the biopsy came out as squamous cell carcinoma of the esophagus. She passed away within a few days before radiotherapy could be commenced.

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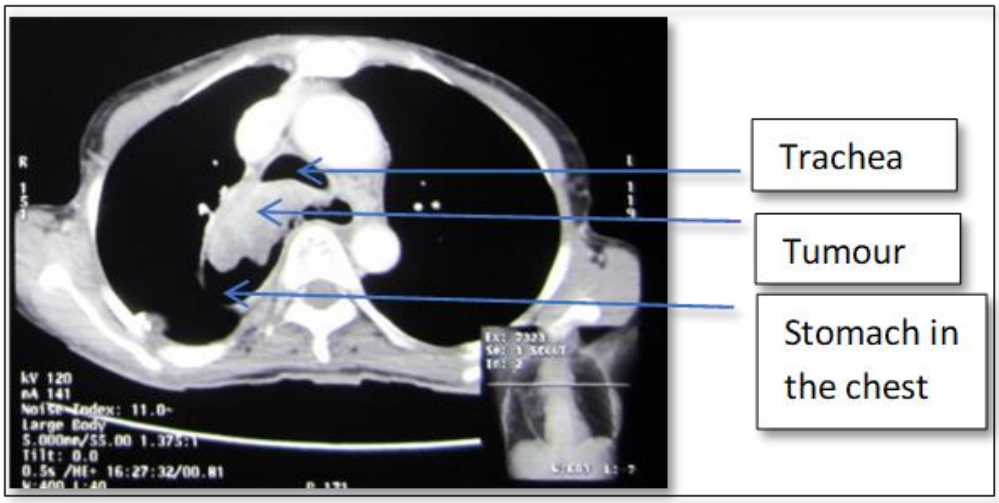

Figure 1. CT scan of the chest showing the tumor which has grown into the stomach and, indenting the trachea.

DISCUSSION

Achalasia of the cardia has long been known as a pre-malignant condition, with the esophageal carcinoma developing several years after the onset of symptoms of the achalasia. Durations varying from 5 to 24 years have been reported [1-4]. 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 esophageal carcinoma 25 years after the initial presentation of the achalasia.

The chronic food stasis over the years affects the distal and mid-esophagus. This leads to irritation of the mucosa, which then leads to metaplasia, dysplasia and finally the carcinoma of the esophagus. Since the food stasis does not usually affect the cervical esophagus, it is often spared from the malignant change. At the second surgery, the patient had transthoracic esophagectomy, leaving behind part of the thoracic esophagus. This is the part that developed the carcinoma several years later. If the esophagectomy had involved the whole of the abdominal and thoracic esophagus, leaving behind only the cervical esophagus, it is unlikely that a malignancy would have developed later, since the cervical esophagus is often not involved with the chronic food stasis and irritation. It is therefore our opinion that when esophagectomy is considered for recurrent achalasia or failed modified Heller's operation, the whole of the thoracic esophagus be removed, and the anastomosis done at the level of the cervical esophagus (with the colon or the stomach), to forestall malignant change in the remnant thoracic esophagus. There must also be long-term surveillance of achalasia patients.

CONCLUSION

Malignant transformation in the esophagus occurs several years after the initial diagnosis of achalasia. When esophagectomy is considered in the course of management of the achalasia, the whole of the thoracic esophagus must be removed and the anastomosis done with the cervical esophagus.

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