Abstract

Achalasia cardia is a known premalignant lesion for esophageal malignancy. Although squamous cell carcinoma is the most common malignancy reported in long standing achalasia patients, few reports of adenocarcinoma due to Barrett’s metaplasia are also published in patients who have undergone pneumatic dilation for achalasia. Patients require either surgery or pneumatic dilation to avoid the complications of long standing achalasia. Esophageal malignancy due to long standing achalasia requires surgery, chemoradiotherapy or palliative therapy depending on the stage of presentation. We report a case of squamous cell carcinoma of esophagus arising in a patient with long standing achalasia.

Introduction

Achalasia is an idiopathic disorder of esophagus involving lower esophageal sphincter (LES), characterized by insufficient relaxation of LES and absence of esophageal peristalsis. Worsening dysphagia is the main complaint. Pneumatic dilation or Heller’s cardiomiotomy with fundoplication are the preferred therapies for achalasia. Long standing achalasia can lead to squamous cell carcinoma of esophagus. The risk for development of esophageal cancer is 0.4-9.2% in achalasia.\(^1\) Even adenocarcinoma of esophagus is reported in achalasia patients who have undergone pneumatic dilation, due to gastroesophageal reflux leading to Barrett’s metaplasia.\(^2\) Since initial symptoms are same in both achalasia and esophageal carcinoma, patients with malignancy due to long standing achalasia are usually diagnosed late and in an advanced stage. Although screening endoscopy in achalasia is still not standardized, screening endoscopy should be initiated after 15 to 20 years of disease duration and then periodic surveillance every two to three years is advised.\(^1\)

Case report

A 58 years female presented with history of dysphagia for the previous 30 years. She was diagnosed as having achalasia cardia at that time and had undergone pneumatic dilation thrice previous to this admission. The last dilation was done 8 years prior. After the last dilation, the patient continued to have some solid and liquid dysphagia and occasional vomiting. At her most recent presentation, she now complained of increase in severity of dysphagia for both solids and liquids. Physical examination did not reveal any positive findings. Barium esophagram revealed an irregular filling defect in lower esophagus along with dilated esophagus both distal and proximal to the filling defect with a “bird’s beak” sign at the esophagogastric junction. (Figure 1) She underwent upper gastrointestinal endoscopy which revealed a 2 cm nodular, friable mass at 28 cm from the incisors without luminal compromise. (Figure 2A & 2B) Multiple biopsies were taken. Her esophagus was dilated, filled with food residue and there was resistance to passage of scope across the gastroesophageal junction, see videos 1 and Figure 3. Histopathology of the mass revealed moderately differentiated squamous cell carcinoma. Computerized tomography (CT) scan showed thickening of lower
2/3rd of esophagus with gross dilation of esophagus and multiple lymph nodes in the mediastinum with largest measuring 1.2 x 1.2 cm. This patient with a long history of primary esophageal achalasia was diagnosed as having metastatic squamous cell carcinoma and referred for chemoradiotherapy.
Discussion

Achalasia as a cause of esophageal cancer was first reported by Fagge in 1872. Since then achalasia has been considered as a premalignant condition for esophageal malignancy. Bolivar and Herendeen observed 0-7.7% of clinically determined malignancy but a higher incidence of 20-29% malignancy at autopsy. Epidemiological data have demonstrated an annual incidence in the range of 0.5 cases per 100,000 people, with a prevalence of about 8 cases per 100,000 people per year. The risk of esophageal malignancy for patients with long-standing achalasia is between 14 to 140 fold higher than the general population. It has been hypothesized that the chronic stasis of food and bacterial overgrowth seen in long standing achalasia is responsible for dysplasia and eventual esophageal carcinoma. Although squamous cell carcinoma is the most commonly associated malignancy, adenocarcinoma in Barrett’s metaplasia has been reported in achalasia patients developing chronic gastroesophageal reflux after pneumatic dilation.

Whether regular pneumatic dilation or cardiomyotomy offers protection from malignancy is not certain since malignancy has been reported in patients who have undergone either pneumatic dilation or cardiomyotomy. While some authors recommend early surgery for prevention of esophageal cancer, the others content that early surgical intervention does not prevent the development of esophageal cancer. This may be because of inadequate treatment and also associated hypoperistalsis in achalasia. Also, once megaesophagus is developed, surgery does not relieve the stasis completely.

Studies have shown an interval of at least 15 years between the first symptom of achalasia and the diagnosis of esophageal carcinoma. However, achalasia is often not recognized early and it is not uncommon that for patients to have dysphagia 10 years or more by the time achalasia is diagnosed. Therefore, patients may be already at risk for esophageal cancer when achalasia is identified. Also both esophageal cancer and achalasia have similar symptoms thereby making the early diagnosis of cancer in achalasia difficult. Large amount of retained food in megaesophagus impairs visualization adding to the problem of early diagnosis. Therefore, the majority presents in advanced stages and around 80% are inoperable at the time of diagnosis. Despite these concerns for increased risk for development of advanced esophageal cancer among achalasia patients, there is no consensus for screening in case of achalasia. However surveillance is recommended in long standing achalasia so that detection at an early stage may offer curative therapy.

The treatment of choice for early stage cancer is surgery. Unfortunately, when esophageal carcinoma arises in the achalasia esophagus, the tumor is large and advanced in stage before symptoms manifest in the already dilated achalasia esophagus. Therefore multimodality treatment is often utilized for better survival in these patients. Recent studies have shown that the combination of radiosensitive chemotherapy and external beam irradiation is superior to radiotherapy alone.

Conclusions

Achalasia is a premalignant lesion for esophageal cancer. Both squamous cell carcinoma and adenocarcinoma can develop in achalasia patients. Patients are usually diagnosed late and multimodality therapy may prolong the survival. Regular pneumatic dilation or cardiomyotomy does not protect from the occurrence of cancer in achalasia. Though endoscopic pneumatic dilation is a universally accepted therapy, there is no consensus for screening in achalasia. Endoscopic surveillance in patients with long standing achalasia may detect cancer in early stage so that patient will be benefitted with curative therapy.
References


