

Letters to the Editor

Advanced esophageal squamous cell carcinoma in patient with longstanding achalasia

Key words: Esophageal achalasia. Squamous cell carcinoma. Risk factor.

Dear Editor,

Achalasia is the most common esophageal motor disorder, with an estimated annual incidence of 1-2 per 200,000 inhabitants (1). It is characterized by a loss of esophageal peristalsis and incomplete relaxation of lower esophageal sphincter, often hypertonic, causing a functional obstruction of esophageal transit (2), cause of the symptoms and complications of the disease. It has been reported the association of longstanding achalasia with esophageal squamous cell carcinoma, with a risk 16 times higher than in the general population (3).

Case report

Male 56 years old, smoker, diagnosed achalasia fifteen years before, performing endoscopic dilatation at that time, without treatment or follow because of a decision by the patient. Enter for a 3-month history of progressive dysphagia to solids and liquids and weight loss. Oral endoscopy was performed, showing a esophagus with abundant fluid, with dilated walls, sigmoid aspect and mucosal aspect cracked, friable and cardia punctate, with resistance to the passage of the endoscope (Fig. 1) without injury under cardia that justified pseudoachalasia. Twenty two centimetres from dental arch, it was observed a excavated mucosal injury, deep, covered with fibrin and blood cell stippling, covering half the circumference and with malignant aspect. Histology was moderately differentiated squamous cell carcinoma

(Fig. 2). CT scan showed a dilated esophagus with thickening of the proximal third esophageal wall and infiltration of tracheal posterior wall (Fig. 3), which is confirmed by bronchoscopy. With the diagnosis of esophageal squamous cell carcinoma stage III, moderately differentiated, with tracheal infiltration, patient was treated with systemic chemotherapy as cisplatino/5-Fluorouracilo scheme. The patient suffers an aspiration episode with progressive clinical deterioration, occurring finally the death seven days after initiation of treatment. This was in conjunction with tracheal perforation, bronchopneumonia and mediastinitis.

Discussion

Functional alterations of achalasia are due to a degenerative-inflammatory process of esophageal myenteric plexus of unknown etiology that leads to the destruction of the neurons responsible for the relaxation of the sphincter and esophageal persistalsis (4).

The onset is insidious, being dysphagia with progressive worsening the main symptom. Regurgitation of food is the sec-



Fig. 1. Evolved achalasia with esophageal dilatation, cracked mucosa distal and cardia punctate.

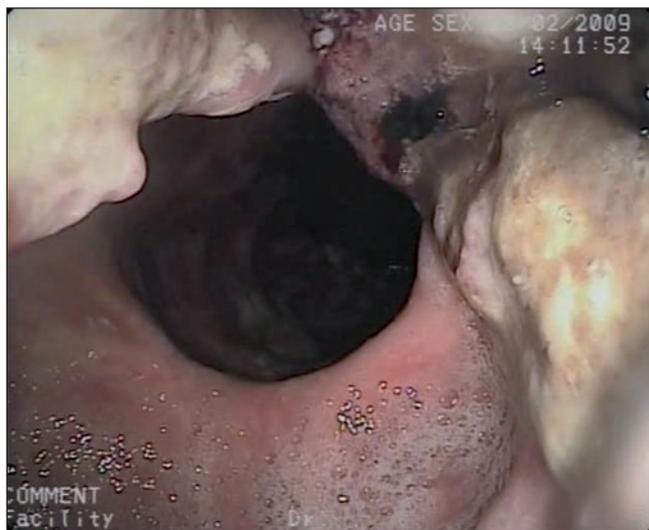


Fig. 2. Esophageal squamous cell carcinoma.

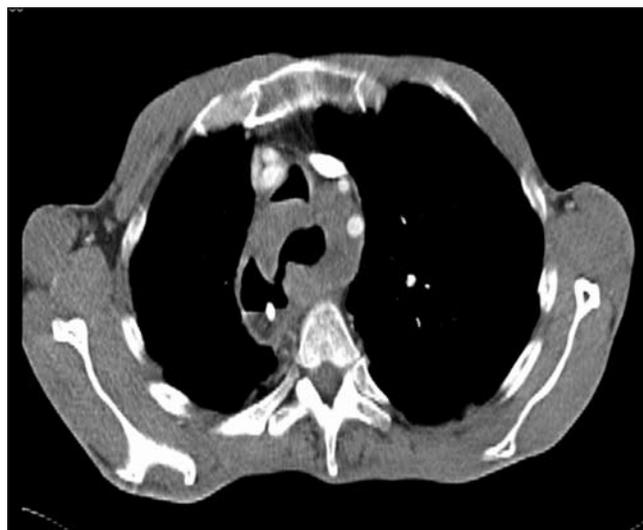


Fig. 3. CT: wall thickening of proximal third of esophagus with infiltration of the posterior tracheal wall.

and in order of frequency and chest pain occurs in one third of patients, being often the cause of reduced intake and loss weight. In advanced stages, there may be passive regurgitation, due to food retention, with recurrent episodes of aspiration and bronchopulmonary complications (5).

With clinical suspicion and a compatible barium study, the diagnosis is confirmed by the performance of esophageal manometry (6), supplemented with endoscopic study, which is essential to detect the presence of neoplasia at the gastroesophageal junction. In advanced cases, in the distal third, the mucosa usually shows a cracked and friable appearance as a result of stasis esophagitis. We can also find changes in the esophageal lumen in cases of megaesophagus. The cardia is closed and provides resistance to the passage of the endoscope.

Treatment is directed at improving symptoms and preventing complications. The most widespread forms are surgical myotomy, pneumatic dilation and botulinum toxin injection. The pneumatic dilation and surgery are alternative treatments, with similar success rates between 75-80%, somewhat higher with surgical treatment. The pneumatic dilation has a better balance of cost-effectiveness, an almost immediate recovery and maintenance of surgery as salvage therapy (7).

Nowadays it is accepted that achalasia is a risk factor for developing squamous cell carcinoma, most frequent complication in patients with ineffective treatment or untreated (8). The food stasis leads to prolonged contact between some elements and the epithelial surface, precipitating factor for this complication. The risk is increased in achalasia of more than 15 years of evolution (9) and tumors usually arise in a widely dilated esophagus. The symptoms may be delayed and when detected, the tumors are usually large and advanced (10).

Although it is not considered a regular monitoring program as a standard practice, there have been periodic endoscopic follow-up recommendations in these patients (11) accounting for achalasia as a premalignant condition, particularly if other risk factors are (such as smoking and alcoholism), as in the case of our patient.

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