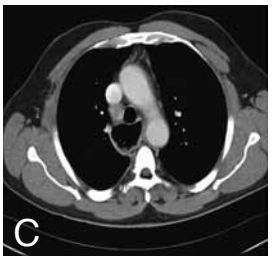
JBR–BTR, 2013, 96: 46.

IMAGES IN CLINICAL RADIOLOGY



Cancer of the oesophagus in achalasia

S. Maeyaert[1], J. Pringot[2], J.L. Samain[1], L. Ghijseling[2], P. Matthys[2]

A 54-year old man was admitted to the emergency department for chronic dysphagia for solid as well for liquids of several years duration, with increasing severity for two weeks.

The patient also had shortness of breath grade III for two weeks.

The medical history of the patient was uneventful, except for surgery on the oesophagus 26 years previously, probably for achalasia. No follow-up was performed. At the time of admission, the patient doesn't take any medication. He doesn't smoke and consumes one or two alcohol units a day.

Clinical examination fails to reveal any specific findings. Blood analysis showed a decrease of the hemoglobin level (10.4 g/dL), a slightly elevated leukocytosis (10400/µL) and elevated C-reactive protein level (127 mg/L), as well as hyponatriemia and hypochloremia.

Chest X-ray shows cardiomegaly and a dilated proximal oesophagus with a marked decrease in the diameter distally.

Fibroscopy shows an ulcerated tumoral lesion of 6 cm at 40 cm, biopsies showing an infiltrating epidermoid carcinoma.

Bronchoscopy was negative. PET-scan confirms the presence of a large, irregular, hypermetabolic mass in the lower oesophagus and enlarged hypermetabolic mediastinal lymph nodes.

Barium X-ray confirms the predominantly enlarged upper oesophagus (4 cm) with stasis of liquids and absence of peristalsis. The distal part of the oesophagus is narrowed showing a pattern compatible with treated achalasia (Fg. A). On the right posterior border of the distal one third of the oesophagus the large ulcerated lesion of approximately 8 cm with typical neoplastic pattern is demonstrated on the lateral view (Fig. B). Contrast enhanced computerized tomography of the thorax confirms the presence of the ulcerated mass and shows an enlarged lymph node in the mediastinum and the right hilus (Fig. C). Treatment consisted of radio-chemotherapy.

Comment

Achalasia is a primary oesphageal motility disorder affecting mainly adults of both sexes equally and predominantly presenting with dysphagia, sometimes with regurgitation.

The treatment of achalasia is symptomatic and designed to improve the oesophageal emptying. The most effective treatments are pneumatic balloon dilatation and cardiomyotomy, follow-up being based on X-ray and manometry. Late complications are related to recurrent dysphagia after treatment. Perihiatal scarring, peptic stricture or obstructing tumors can all cause recurrent symptoms. The most important late complication is the occurrence of oesophageal cancer, patients having similar symptoms in recurring achalasia and in oesophageal cancer. Therefore, mostly cancers are diagnosed in an advanced stage.

The relationship between achalasia and cancer was first described in 1872 by Fagge.

An increased risk of developing oesophageal carcinoma in patients with long standing achalasia has been reported by several authors, with incidences varying widely from 1.7 to 20%.

It has been established that the consumption of alcohol and tobacco increases the risk of developing esophageal cancers due to the content of carcinogens in alcohol and tobacco. Achalasia is known to cause retention and increased bacterial growth and chemical irritation from the continuous composition of food that culminates in chronic oesophagitis. This is supposed to provoke cancer or make the epithelium more sensitive to carcinogens.

In achalasia, either adenocarcinoma or squamous cell carcinoma may develop.

Squamous cell carcinoma is the most frequent (85% of the cases) and situated in over half the cases in the middle third of the esophagus due to stasis. Adenocarcinomas are most often situated in the distal third of the oesophagus due to Barett oesophagitis, consequent to gastrointestinal reflux.

Due to the localisation in the enlarged, dilated oesophagus, tumors are mostly very large at time of diagnosis.

The classical imaging technique for following treated achalasia and recurrence or complications on the oesophagus is barium X-ray study. As demonstrated in the reported case, it is important to examine the entire oesophagus in many incidences to investigate the whole circumference of the dilated oesophagus. Otherwise, even an advanced lesion may remain undetected. In this example, the very large carcinoma is poorly visible on the AP incidence, but well demonstrated on the lateral view.

The ulceriform cancer of the oesophagus is characterized by an ulceration surrounded by a ridge that bulges into the oesophageal lumen. In profile, the lesion creates an image of an oval niche underlined by a radiolucent band. Depending on the stage of development of the cancer, the opposite wall either remains pliable with mucosal folds, or is rigid, without any mucosal relief.

Prognosis of cancers associated with achalasia is very poor due to the late stage of diagnosis. Early treatment of achalasia relieving the retention reduces the risk of oesophageal cancer. However, cancer risk continues after treatment so endoscopic or radiological follow-up is necessary, even after treatment.

Reference

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1. Department of Radiology, University Hospital Leuven, Leuven, Belgium, 2. Department of Radiology, Sint Elisabeth Hospital, Brussels, Belgium.