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Adenosquamous carcinoma of the esophagogastric junction. Case report

F. FRANCIONI, S. TSAGKAROPOULOS, V. TELHA, R. BARILE LA RAIA, T. DE GIACOMO

SUMMARY: **Adenosquamous carcinoma of the esophagogastric junction. Case report.**

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Adenosquamous carcinoma is a rare tumor with coexisting elements of infiltrating squamous cell carcinoma and adenocarcinoma. This tumor is reported to arise in different organs but rarely in the oesophagus. In most cases, it shows highly aggressive biological behaviour with high propensity to regional lymph-node metastasis and poor prognosis. We describe the management of a patient with an aggressive adenosquamous carcinoma of the esophagogastric junction.

RIASSUNTO: **Carcinoma adenosquamoso della giunzione esofago-gastrica. Descrizione di un caso.**

F. FRANCIONI, S. TSAGKAROPOULOS, V. TELHA, R. BARILE LA RAIA, T. DE GIACOMO

Il carcinoma adenosquamoso rappresenta una rara varietà istologica con i caratteri misti del carcinoma squamoso e dell'adenocarcinoma. Questo istotipo si presenta in diversi organi ma raramente a livello esofageo. In questi pazienti la prognosi è sfavorevole per l' elevata aggressività biologica del tumore e la tendenza alle metastasi nei linfonodi loco regionali. Descriviamo il caso di un paziente con diagnosi istologica di carcinoma adenosquamoso della giunzione esofago-gastrica sottoposto ad intervento di esofagectomia.

KEY WORDS: Esophageal carcinoma - Adenosquamous carcinoma - Esophagectomy - Gastric tube - Esophagogastric junction. Carcinoma dell'esofago - Carcinoma adenosquamoso - Esofagectomia - Tubulo gastrico - Giunzione esofago-gastrica.

Introduction

Adenosquamous carcinoma (ASC) of the esophagus is an uncommon histotype of esophageal cancer. Few cases have been described in the literature. In this type of cancer, elements of squamous cell carcinoma (SCC) and adenocarcinoma (ACE) coexist (1). Usually ASCs, because of their highly aggressive biological behavior and their high propensity to regional lymph nodes metastasis (2), are usually associated with worst prognosis than the common histotype of esophageal cancer. Since ASC are rare tumors, the real incidence and the clinicopathologic features are not fully characterized. Most cases described in the literature are localized in the middle esophagus (3).

We describe the case of an aggressive ASC in the lower third of the esophagus involving the esophagogastric junction.

Case report

A 75-year-old white man was admitted at our Department with a 3-months history of dysphasia and weight loss. He also had a 40 pack/year history of cigarette smoking. The barium swallow study showed a filling defect in the distal esophagus, above the esophagogastric junction. At the endoscopic control, a partially ulcerated protruding mass with infiltrative aspect was revealed at the lower third of the esophagus. Multiple biopsies were obtained and specimen showed the presence of a mild-differentiate squamous cell carcinoma. Whole body Computed Tomography confirmed the esophageal tumor with 5 cm longitudinal extension at the esophagogastric junction, with no evidence of mediastinal lymph nodes or distant metastases. Routine preoperative investigations demonstrated reduction of HGB (10.1 mg/100ml), RBC (3.7 million/mm3), HCT (30.5 %)

"Sapienza" University of Rome, Italy
"P. Stefanini" Department of Surgery and Transplant, Thoracic Surgery

and PLT (147 thousand /mm3). Pulmonary function tests indicated a 2.26 L of FEV1. Total esophagectomy was performed through a right thoracotomy; the tumor was removed ''en bloc". Extended lymphadenectomy of the mediastinum and abdominal compartment was completed. Continuity of alimentary tract was re-established by interposition of a "narrow" gastric tube placed in the posterior mediastinum and left-side cervical anastomosis.

The patient had an uneventful postoperative course. On the seventh post-operative day, the water-soluble contrast swallow demonstrated regular draining through the gastric tube. Histology showed an ulcerative and infiltrative adenosquamous carcinoma of the esophagogastric junction, classified as type 3 according to the guidelines for clinical and pathologic studies of carcinomas of the esophagus established by the Japanese Society for Esophageal Diseases (Fig. 1) (4). Definitive staging was pT3N1M0, stage III. There was'nt normal mucosa between the two histotypes; that confirmed the non syn-

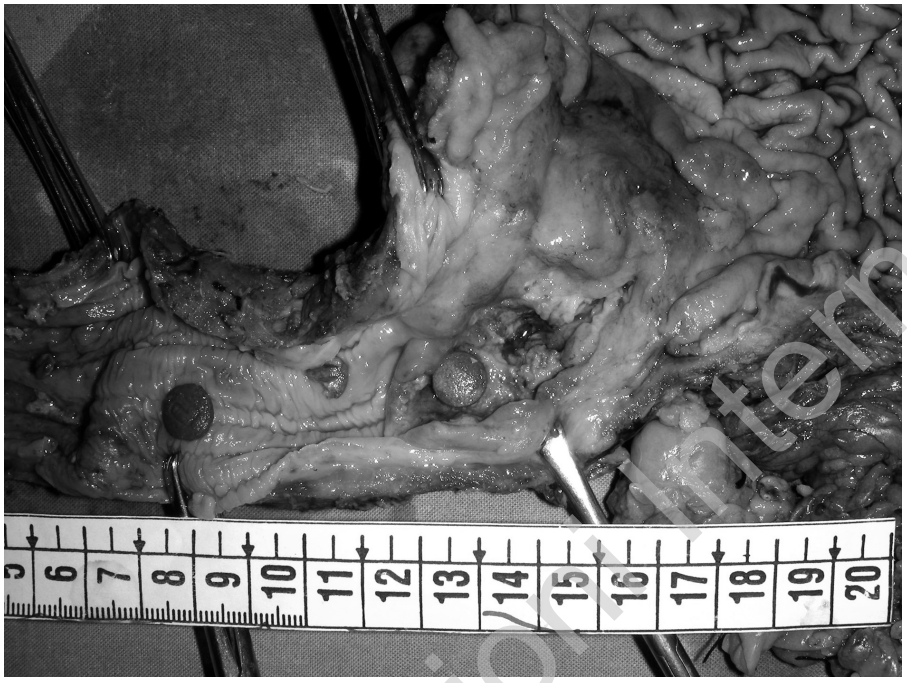

Fig. 1 - Gross appearance of the surgical specimen. A shallow neoplastic ulcer is evident; two pills in the esophageal lumen are also visible.

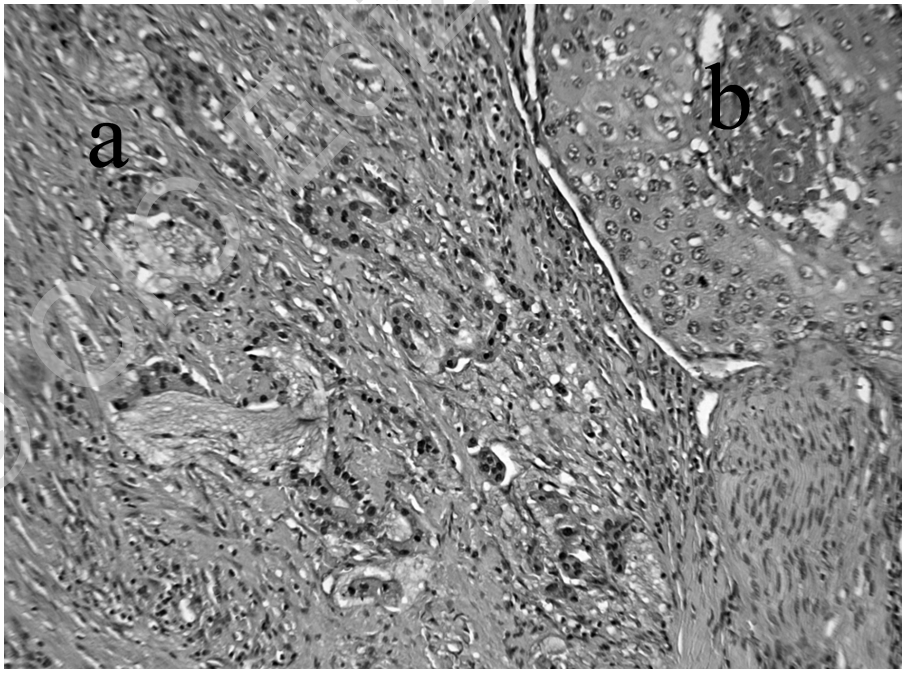

Fig. 2 - Typical microscopic histological features of an ASC of the esophagus: histologic transition between the squamous cell carcinoma (a) and adenocarcinoma components (b) is well evident in the deep part of the tumor.

chronous nature of this tumor according to the Waren and Gates criteria (5).

The patient went through adjuvant chemo-radiotherapy. The disease-free survival was 12 months; the patient died of mediastinal recurrence and distal metastases twenty-four months after surgery.

Discussion

ASC of the esophagus is a rare subtype of esophageal carcinoma containing coexisting elements of infiltrating AC and SCC. According to the guidelines for clinical and pathological studies of carcinomas of the esophagus established by the Japanese Society for Esophageal Diseases (4), ASC is defined as having at least 20% each of SCC and AC elements on routine microscopic examination with hematoxylin and eosin staining. The World Health Organization classification however states simply that ASC has a significant squamous cell carcinoma component that is intermingled with tubular adenocarcinoma elements, with no special reference to the ratio of these two components (6).

There are no clinicopathological features, such as symptoms, tumour markers, location and macroscopic aspects, characterizing this histotype. The use of increased serum concentrations of squamous-cell-related antigen and carcinoembryonic antigen as specific marker is yet to be demonstrated (7, 8). Preoperative endoscopic biopsies often fail to determine the dual nature of the tumor. Primary ASCs have been reported to arise in different organs. In most cases, these tumours show highly aggressive biological behaviour with high propensity to regional lymph node metastasis and poor prognosis.

Regarding the histogenesis of esophageal ASCs, several hypothesis have been proposed based on experimental studies and clinical reports. It has been speculated that the AC elements originates from esophageal glands or their ducts. However, studies demonstrated neither cellular atypia nor transition to tumor cells in the esophageal glands and their ducts (9). This hypothesis is also contradicted by the fact that ASCs of the esophagus can be induced in rats, which have no submucosal esophageal glands. In addition Steele and Nettesheim have demonstrated that a single cell isolated from mixed adenosquamous cell carcinoma in rats can differentiate into pure adenocarcinoma, squamous cell carcinoma, or composite tumor consisting of both histologic types through differentiative instability of single-cell clones (10).

In conclusion, ASC carcinoma is a rare tumour with no specific definition of its pathogenesis. Histological preoperative diagnosis is often difficult or impossible and

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