Letter to the Editor

Carcinosarcoma of the esophago-gastric junction—a case description

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Introduction

Carcinosarcoma was firstly described by Virchow in 1865 (1). When seen in the esophagus it is most often localized (80%) in the thoracic portion just below the bifurcation of the trachea (2). Esophageal carcinosarcoma (EC) is usually diagnosed at an earlier stage than esophageal squamous cell carcinoma (SCC) and is commonly restricted to the wall of the esophagus. The therapy is essentially the same as the therapeutic protocol for SCC or adenocarcinoma of the esophago-gastric junction (AEG) of the esophagus (3).

We present a case of resected EC with an atypical location below the esophageal-gastric junction (EGJ) which posed a threat to the patient's life. To our knowledge, this is the only case with such an atypical location for this tumor.

Case description

Medical history

A 68-year-old man was admitted to John Paul's II Hospital because of upper gastrointestinal tract bleeding. Other symptoms, such as weakness, weight loss of approx. 7 kg, accompanied the patient for approx. 2–3 months. After laboratory analyses, severe anemia was found and treated by transfusion of 4 units of packed red blood cells (PRBC). The patient's medical history included: partial resection of the stomach due to peptic ulcer disease 35 years ago, two strokes, and an epidural hematoma.

Imaging examination

Endoscopy was performed at admission and revealed a bleeding polypoid polyp descending from cardia into the stomach. On day 5, follow-up gastroscopy was performed for a precise clinical evaluation, which was impossible on the first examination. During gastroscopy the following findings were present: esophagitis just above the gastric cardia, a 4 cm partially ulcerated polyp descending from the cardia into the stomach (foto1) post-operative findings consistent with previous partial gastrectomy i.e., Rydygier resection, and chronic gastritis. Biopsy specimens were performed from the polypoid mass.

After the diagnosis of cancer, a chest CT scan and positron emission tomography scan (PET-CT) was performed (*Figure 1*). The imaging procedures did not reveal any distant metastases.

Preoperative diagnosis

Histopathological examination of the specimens from the gastric biopsy established the diagnosis of cancer suggestive of EC. The preoperative diagnosis was classified as T2N0M0, stage IIA (4).

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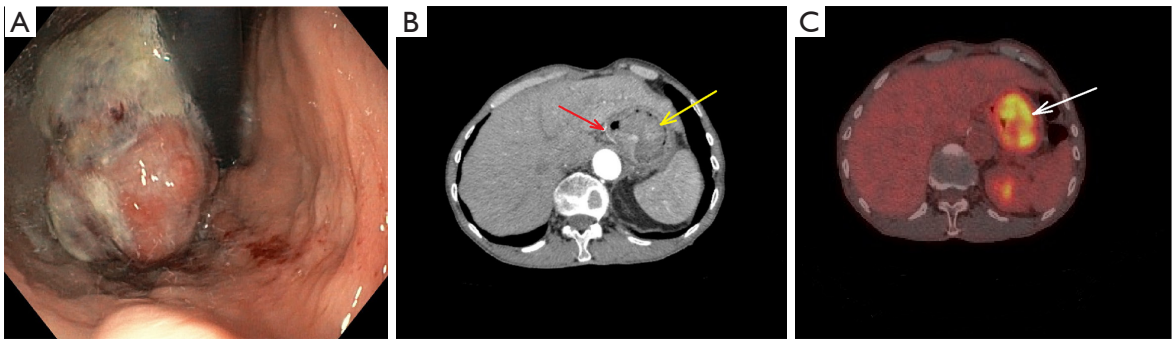

Figure 1 Diagnostic images in a 68-year-old man with carcinosarcoma of the esophago-gastric junction. (A) Gastroscopy. Ulcerated polyp located in the subcardial area under reflected endoscopy. (B) Computer tomography of tumor located below cardia (red and yellow arrows). (C) Positron emission tomography scan (PET-CT) images with tumor showing standardized uptake (arrow).

Treatment

Following multidisciplinary consultation with an oncologist, radiotherapist, and surgeon the patient was qualified for surgery. Due to two strokes, atherosclerosis the patient did not qualify for pre- and post-surgical chemotherapy and radiotherapy. Using an abdominal approach, a gastrectomy including the abdominal part of the esophagus and a D2 lymphadenectomy was performed. For restoration of GI-tract continuity, a Roux-en-Y esophagojejunostomy without formation jejunai pouch, and jejunojejunal anastomosis ca 40 cm from anastomosis was performed (5). The post-surgical period was uneventful. The patient was discharged home on the 10th postoperative day orally nourished with good tolerance.

Outcome (Figure 2)

(I) Gastric biopsy specimens (No. 250997) encompassed five small pieces of the malignant neoplasm with a completely ulcerated surface. The neoplasm was composed of pleomorphic large cells. By immunohistochemistry, the cells did not express panCK (CKMNF116), S-100 protein, LCA, CD34, or smooth muscle actin (SMA). The differential diagnosis established was cancer with characteristics of sarcoma, GIST, or sarcomatous carcinoma.

(II) The surgical specimen (No. 253237) sent to the laboratory included an approximately 90 mm segment of the distal part of the esophagus and stomach, a small amount of fatty tissue from the gastric curvatures, and lymph nodes from groups N1 1–6 and N2 7–11. The polypoid mass, 49 mm in diameter, was present 2 cm below the proximal esophageal resection line anchored by a stalk to the mucosa. The slides obtained from various areas of the ulcerated tumor revealed the presence of two malignant components: a poorly differentiated, non-keratinizing squamous carcinoma (with expression of CKMNF116; not present in the gastric biopsy specimens) and an undifferentiated high-grade sarcoma lacking CKMNF116 expression (*Figure 2F*). The cancer was partially ulcerated with areas covered by squamous epithelium indicating it originated from the esophagus. The carcinosarcoma infiltrated the muscularis propria but was restricted to the esophageal wall. The surgical margins (2.2 cm) and the additional esophageal ring (with a width of 8–10 mm) were negative for carcinosarcoma and all twelve lymph nodes were without metastases. The final classification was pT2N0M0, stage IIA (4).

Follow-up

Patient in the post-operative course was subject to check-ups in the first year every 3 months and in subsequent years every 6 months. During the follow-up, images were routinely taken of the patient's chest examination, chest and abdominal tomography (twice a year), gastroscopy (twice a year). Gastroscopy and a CT scan showed no signs of neoplastic disease recurrence. Despite the accompanying neurological burdens, a good result of surgical treatment was obtained. The patient was on a full diet intake, but without weight gain, BMI remained at the level of about 18. The patient lived for 38 months after surgery and died of a third stroke.

All procedures performed in this study were in

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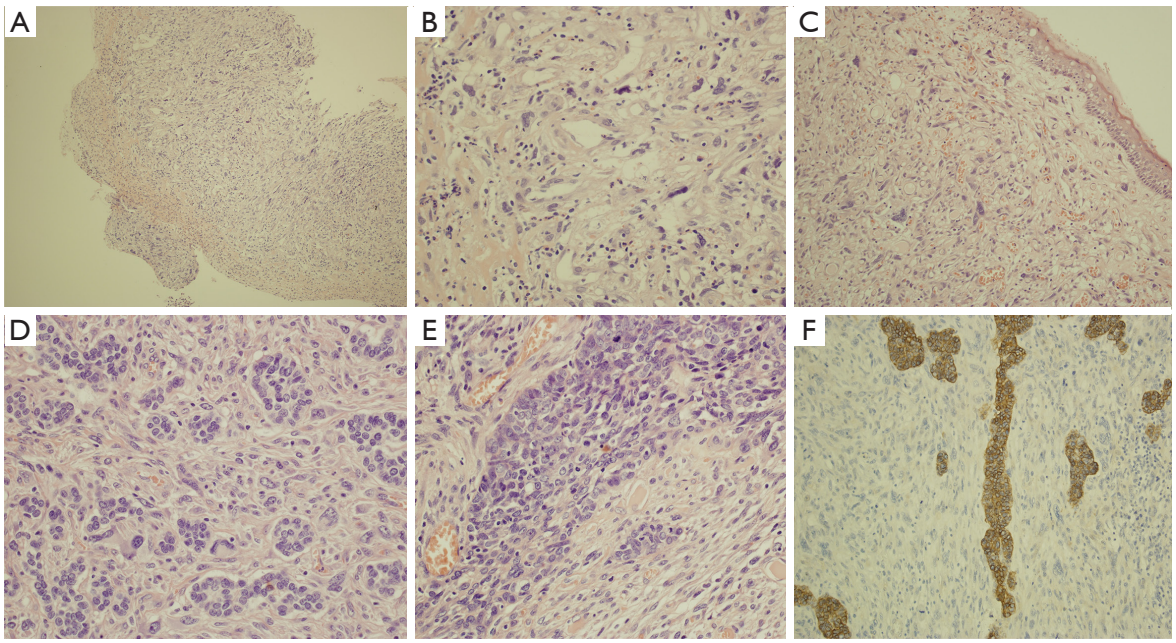

Figure 2 Histopathologic texture of the carcinosarcoma located in the esophago-gastric junction. (A) Gastric biopsy specimen with ulcerated surface showing complete infiltration by a neoplastic sarcoma. HE staining. Objective magnification ×10. (B) Gastric biopsy specimen with high-grade undifferentiated sarcoma composed of pleomorphic cells lacking any carcinoma component. HE staining. Objective magnification ×40. (C) Surgical specimen showing superficial part of the polypoid esophageal tumor occupied by the sarcomatous infiltrate covered by esophageal squamous epithelium. HE staining. Objective magnification ×20. (D) Surgical specimen showing the deep part of the tumor with sarcoma and carcinoma components. HE staining. Objective magnification ×40. (E) A more distinct squamous carcinoma component of the esophageal tumor from the surgical specimen with a poorly visible sarcomatous component. HE staining. Objective magnification ×40. (F) Immunohistochemistry of the surgical specimen—cytokeratin (CKMNF116) positivity in the carcinoma and lack of its expression in the sarcoma area. Objective magnification ×20.

accordance with the ethical standards of the institutional and/or national research committee(s) and with Declaration of Helsinki (2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

Carcinosarcoma is a rare biphasic tumor. In the available literature, approximately 130 cases in the esophagus and 60 in the stomach have been reported. Grossly it presents as a polypoid mass (75%), however, it can present as an ulcerative infiltrate (25%). When sarcoma is the dominant component the tumor takes a polypoid form, and when the carcinomatous component is dominant the tumor displays an ulcerative growth pattern (6). The incidence of such tumors has been reported to be 0.4 to 2.4% among all esophageal neoplasms (7). In about 80% of cases, EC is confined to the esophageal wall and displays a course less aggressive than that of SCC (3). Matsusaka *et al.* divided sarcomatous esophageal tumors into 3 groups: (I) carcinosarcoma, (II) pseudosarcoma, and (III) true carcinosarcoma (8). Ro *et al.* proposed the histopathological criteria for carcinosarcoma: the simultaneous presence of malignant epithelial and spindle cell components separated by transitional areas and the sarcomatous component expressing a distinct epithelial phenotype (9). The Japanese Society of Esophageal Carcinoma postulated three theories referring to carcinosarcoma histogenesis. The first theory suggests these tumors occur as a spindle cell reaction in response to a developing carcinoma. The second theory suggests these tumors develop from the simultaneous development of two different cancers from two stem cells. The third theory suggests a metaplastic pathway leading to a sarcomatous component from epithelial cells (3).

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It is suggested that in most cases carcinosarcoma contains a sarcomatous component as a result of SCC differentiation. The basis for this theory is the presence of a transitional zone between the carcinomatous and sarcomatous components in cases of the most commonly occurring type, carcinosarcoma. The true type is much less common (3,10). Molecular analysis points out the differences between the two components of carcinosarcoma. The molecular aberrations affecting the P53, cyclin, D1, MDM2, and CDK4 genes were examined (11,12). Immunohistochemistry is helpful in the diagnosis of EC. Cytokeratin expression identifies carcinomatous differentiation, whereas vimentin positivity, although nonspecific, points to the mesenchymal origin of spindle cells (2,12). Despite the diagnostic possibilities, the histopathological entity and biology of the tumor remain controversial. This is confirmed by new reports describing only three cases of adenocarcinosarcoma at the EGJ so far (13-15). Our report presents the first case of EC with a squamous component in a location below the EGJ.

Currently, no guidelines are available for the treatment of EC thus the protocols for esophageal SCC and AEG are used. The procedure of choice is subtotal esophageal resection with a 2 or 3 field lymphadenectomy if located in the thoracic portion, and gastrectomy with D2 lymphadenectomy for abdominal locations with preoperative chemo-radiotherapy. However, due to the sarcomatous component, the use of chemo- and radiotherapy is recommended (3,13-15). Despite EC being less advanced at the time of diagnosis than SCC or AEG, the long-term prognosis is not much better (13,14). The three-year survival is better for EC than for SCC (63% *vs.* 28%, respectively) but the five-year survival rate is comparable (27% *vs.* 22%) (15).

In conclusion, a sarcomatous component of cancer at the EGJ is very rare and the management is difficult requiring individualization of treatment protocols.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-22-168/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with Declaration of Helsinki (2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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