Case Report

Atypical Carcinoid of the Esophagus: Report of a Case

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We present a case of an atypical carcinoid in the midthoracic esophagus. Esophagectomy was performed with a three-incisional approach. Although both postoperative chemotherapy and radiation therapy were given, the patient died 11 months postoperatively of liver metastasis and cachexia. We suggest that the aggressive behavior of atypical carcinoid of the esophagus precludes the use of limited surgical resection and requires adjuvant chemoradiotherapy. (Ann Thorac Cardiovasc Surg 2002; 8: 302–5)

Key words: esophagus, neuroendocrine tumors, atypical carcinoid

Introduction

The most common histological type of esophageal tumor is squamous cell carcinoma. Neuroendocrine tumors are exceedingly rare. Atypical carcinoid (AC) is an intermediate form of tumor between low-grade malignant typical carcinoid (TC) and high-grade malignant small cell carcinoma (SCC), which represent the two ends of the spectrum of neuroendocrine tumors. Here we report a case of AC in the midthoracic esophagus.

Case Report

A 54-year-old man presented with a 3-month history of progressive dysphagia and 3-kg weight loss. At the time of presentation he was only able to swallow liquids. No clinical features of carcinoid syndrome were present. His past history and that of his family were not contributory. Physical examination revealed no abnormality. The supraclavicular lymph node was not palpable. Routine examinations of blood, urine and feces gave normal results. A plain chest roentgenogram showed no abnormal shadow apart from emphysema. A barium swallow showed a filling defect about 5 cm in the midthoracic esophagus (Fig. 1). Esophagogastroscopy revealed an irregular intraluminal mass with ulceration localized in the middle esophagus 30 cm from the incisors, partially obstructing the lumen. Investigations in search of possible extrathoracic metastasis, including brain and abdominal computed tomographic scan, demonstrated no cancer.

The patient underwent an esophagectomy. The surgical approach was through combined abdominal incision, right thoracotomy, and left cervical exposure for esophagectomy esophagogastric anastomosis. In operation, the three-field lymph nodes including the cervical (left supraclavicular regions), mediastinal (periesophagus and around the trachea), and abdominal (perigastric region) lymph nodes were dissected.

Grossly, the tumor with focal ulceration measured 5.5×3.0×2.0 cm. Histological examination revealed that the tumor cells were arranged in a nested, trabecular, or pseudorosette pattern, with increased numbers of mitoses, nuclear pleomorphism, and presence of necrosis (Fig. 2). The tumor cells had invaded the full thickness of the esophageal wall. Three of seven lymph nodes examined showed metastases. Immunohistochemical stains were performed and the results showed the tumor cells were positive for neuron-specific enolase (NSE) (Fig. 3) and chromogranin A (CgA) (Fig. 4). The final pathology report was an AC of the esophagus.

The postoperative course was uneventful. The patient was discharged from the hospital 10 days after the operation. Both postoperative chemotherapy (two courses) and radiation therapy (one course) were given. The combination chemotherapy consisted of cis-diamminedichloroplatinum (CDDP) 80 mg/m^2 on day

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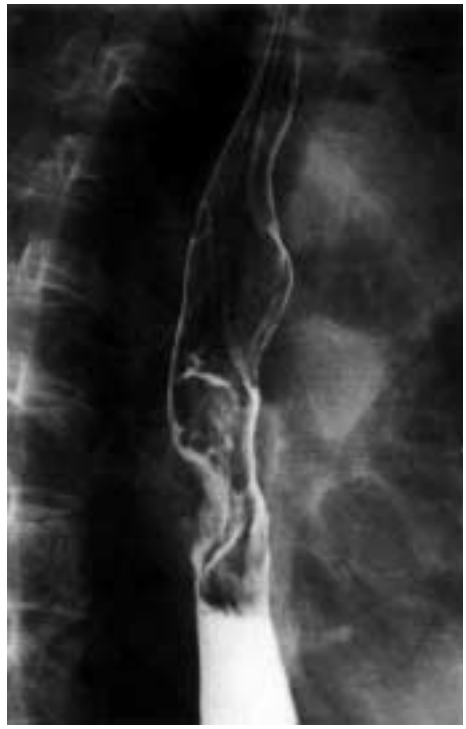

Fig. 1. A barium swallow showed a filling defect about 5 cm in the midthoracic esophagus.

1 and 70 mg/m^2 of etoposide (VP-16) on days 1-4, and the whole irradiation of the thoraxes was 60 Gy. The patient died 11 months postoperatively of liver metastasis and cachexia.

Discussion

Carcinoid tumors are neuroendocrine tumors and are thought to arise from amine precursor uptake and decarboxylation (APUD) cells.[1] Because the distribution of APUD cells in the esophagus is sparse compared to that in the intestine and appendix, esophageal carcinoid tumors are exceedingly rare.

Esophageal carcinoid shows no clinical specificity. All the reported cases presented with dysphagia, and only one had the carcinoid syndrome.[2-12] So it is impossible to differentiate esophageal carcinoid tumors clinically from squamous cell carcinoma and adenocarcinoma. The final diagnosis depends on the light microscopic, electron microscopic and immunohistochemical features.

Because AC is a neuroendocrine neoplasm which aggressive ability is intermediate between TC and SCC,[13] it is necessary to differentiate AC from other tumors. Now histological appearance of an AC is established according to the criteria proposed by Arrigoni and associates:[14] 1) increased mitotic activity in presence of recognizable carcinoid pattern; 2) pleomorphism, nuclear irregularity, hyperchromatism, and abnormal nuclear-cytoplasmic ratio; 3) hypercellularity with disorganization of the architecture; 4) areas of tumor necroses. In agreement with Lindberg et al.,[12] we can state that distinguishing AC from poorly differentiated carcinoma may be difficult sometimes due to their analogy in cellular morphology. At this time, argyrophilic staining techniques and the use of electron microscopy to demonstrate neurosecretory granules are helpful for confirming the neuroendocrine nature of

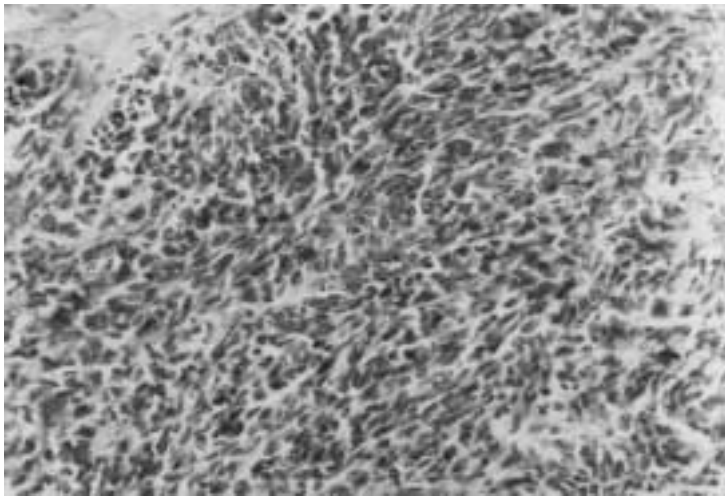

Fig. 2. Photomicrograph showing the tumor cells and nuclei were spindle shaped. (HE staining, ×400)

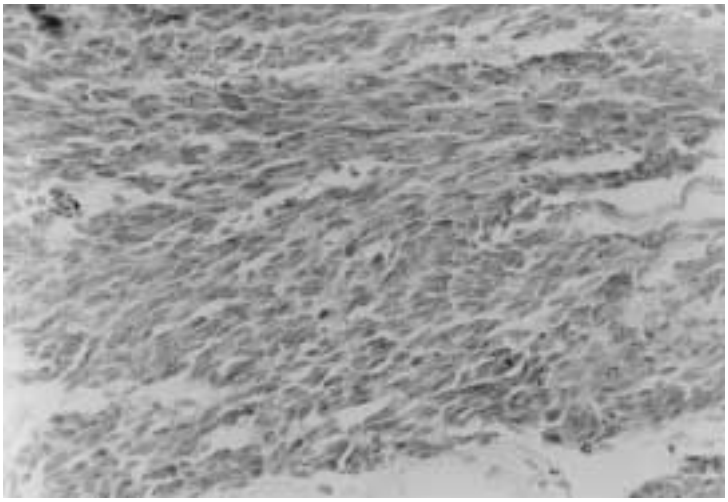

Fig. 3. Shows NSE positive reaction in cytoplasm of the tumor cells. (SABC, ×400)

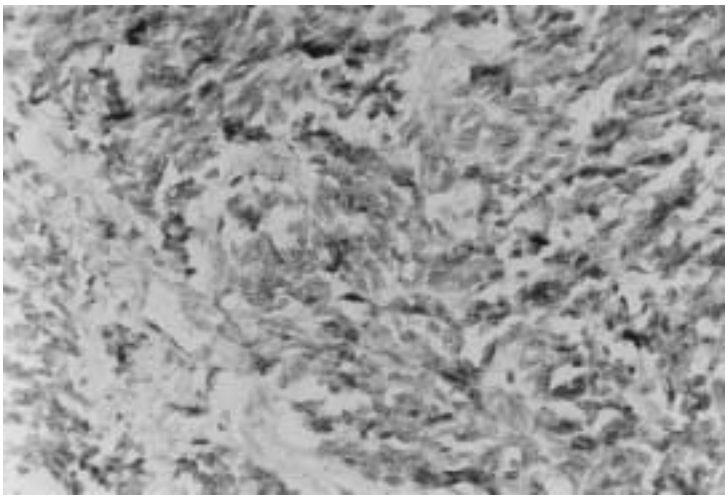

Fig. 4. Shows CgA positive reaction in cytoplasm of the tumor cells. (SABC, ×400)

tumors and thus excluding other esophageal tumors such as squamous cell carcinoma and adenocarcinoma. In addition, immunocytochemical staining, for example, CgA and NSE are good markers for tumors with neuroendocrine features, but are not useful for accurately identifying the neuroendocrine tumor subtypes.

Due to the small number of cases reported so far, the optimum management of AC tumors of the esophagus is unclear. Lindberg et al.[12] reported a case of an upper esophageal AC treated by polypectomy, the patient was free of disease nine months after excision. But considering that the histological and clinical characteristics of AC of the esophagus are similar to those of the lung, we suggest that AC of the esophagus be treated by the same regimen used for AC of the lung. Firstly, radical esophagectomy with systematic mediastinal lymph nodes dissection must be routinely performed to ensure accurate staging and adequate tumor management; secondly, the patient should be treated with a combination of radiotherapy and chemotherapy postoperatively unless the tumor stage classification is stage I.

In our case, although he received adjuvant radiotherapy and chemotherapy, the patient died of liver metastasis at 11 months. In another case reported by Oz et al.,[10] the patient died of recurrence of esophageal AC at 20 months. In our opinion, there is a high possibility of local recurrence and distant metastasis, as in AC of the lung, and the prognosis of AC of the esophagus may be not favorable.

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